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Multiple Jejunoileal Atresiae in a Term Neonate Managed by Single-Stage Primary Anastomosis: A Rare Case from Syria

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Case Report

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Abstract

Introduction and importance:

Jejunoileal atresia (JIA) is among the most frequent causes of congenital neonatal intestinal obstruction, with an estimated incidence of approximately 1 per 10,000 live births. It is generally attributed to a late intrauterine mesenteric vascular accident and is classified into four anatomical types according to the Grosfeld modification of the Louw classification. Type IV atresia, characterized by multiple atretic segments, is uncommon and presents distinct surgical challenges, particularly when occurring in isolation, without associated anomalies. This report discusses a rare case of isolated Type IV JIA in a newborn treated with primary end-to-end anastomosis.

Case presentation:

We report a term Arab male neonate who presented on day 3 of life with bilious vomiting, progressive abdominal distension, and delayed passage of meconium. Plain abdominal radiography demonstrated a triple-bubble sign, and abdominal ultrasonography excluded malrotation and intussusception. After resuscitation fluid, exploratory laparotomy revealed six discrete jejunoileal atretic segments. Resection of approximately 30 cm of small bowel and a single-stage primary end-to-end anastomosis were performed. The postoperative course was uneventful; at the 10-month follow-up, the infant was thriving (weight 9 kg) without features of short-bowel syndrome, anastomotic stricture, or adhesive obstruction.

Conclusion

Multiple isolated jejunoileal atresiae represent a rare entity that requires prompt diagnosis and individualised surgical management. When anatomy permits, en bloc resection with primary anastomosis is a safe and effective strategy. Long-term surveillance for nutritional, growth, and surgical complications remains essential.

Background

Jejunoileal atresia (JIA) is one of the most common congenital causes of mechanical intestinal obstruction in the neonate, with a reported incidence of approximately 1 per 10,000 live births [1]. The accepted pathogenetic mechanism, originally proposed by Louw and Barnard in 1955, is a localised late intrauterine mesenteric vascular insult resulting in segmental ischaemia, necrosis, and resorption of the affected bowel; subsequent work has shown that intrauterine intussusception, segmental volvulus, intestinal perforation, and thromboembolic events may produce a similar morphological outcome (Fig. 5A) [2, 3, 7].

The Grosfeld modification of the Louw–Barnard classification recognises four principal types: Type I (intraluminal membrane with intact bowel wall and mesentery); Type II (atretic segments connected by a fibrous cord); Type IIIa (complete separation with a V-shaped mesenteric defect); Type IIIb (the so-called "apple-peel" or "Christmas-tree" deformity, with the distal bowel coiling around a single perfusing vessel); and Type IV (multiple atretic segments) (Fig. 5B) [4, 8, 9]. Type IV is reported in approximately 6–32% of large series and is frequently, but not invariably, associated with additional anomalies, most commonly cardiac defects, gastroschisis, malrotation, and cystic fibrosis [2, 10, 12].

Antenatal sonographic findings of polyhydramnios, dilated bowel loops, and ascites may suggest the diagnosis in up to half of cases. Postnatally, the cardinal triad consists of bilious emesis, progressive abdominal distension, and failure to pass meconium within the first 24–48 hours of life [2, 3]. Plain abdominal radiography typically demonstrates dilated bowel loops with multiple air-fluid levels; the level and pattern of obstruction may be inferred from the number of dilated loops ("triple-bubble" or more numerous bubbles indicating a more proximal or multiple-level obstruction) [7, 10].

Surgical management remains the cornerstone of treatment. The choice between resection with primary anastomosis, multiple anastomoses, tapering enteroplasty, and staged procedures with enterostomy is dictated by intra-operative findings, residual bowel length, and the patient's haemodynamic status [5, 6, 11]. We present a rare case of isolated Type IV jejunoileal atresia involving six atretic segments, successfully managed by single-stage en bloc resection and primary end-to-end anastomosis. The case is reported in line with the CARE 2013 guidelines.

Case Presentation

Patient information and presenting complaints

A 3-day-old male neonate of Arab ethnicity was referred to our institution with a 24-hour history of bilious (greenish) vomiting following each breastfeed (Fig. 1), progressive abdominal distension, and absence of meconium passage since birth. The infant had been delivered at term (39 weeks of gestation) by elective caesarean section, with a birth weight of 3,500 g and uneventful neonatal adaptation. He had been breastfed exclusively and discharged home with his mother on the first postnatal day.

The pregnancy had been monitored regularly; routine antenatal ultrasonography was reportedly unremarkable, with no documented polyhydramnios, dilated fetal bowel loops, or ascites. There was no consanguinity, no family history of congenital gastrointestinal anomalies, and no parental exposure to teratogens.

Clinical findings on admission

On admission, the infant appeared unwell and mildly lethargic, with clinical evidence of moderate dehydration (depressed anterior fontanelle, dry mucous membranes, and decreased skin turgor). Vital

signs were as follows: heart rate 130 beats/min, respiratory rate 32 breaths/min, peripheral oxygen saturation 98% on room air, and afebrile. The abdomen was uniformly distended, soft, non-tender, and tympanic to percussion, without palpable masses or visible peristalsis. The anal opening was patent, and digital examination yielded no meconium. The remainder of the systemic examination, including cardiac auscultation, was unremarkable.

Diagnostic assessment

Initial laboratory investigations revealed a leucocytosis of 16.8 × 10^9/L with neutrophil predominance, a C-reactive protein of 32.5 mg/L, blood urea of 92 mg/dL, serum potassium of 5.3 mmol/L, mild thrombocytopenia (52 × 10^9/L), and a haemoglobin level corresponding to a red cell count of 3.9 × 10^{12}/L. Coagulation parameters (prothrombin time, activated partial thromboplastin time, and international normalised ratio) were within the reference range for age. The biochemical and haematological derangements were considered to reflect prerenal azotaemia secondary to dehydration and a systemic inflammatory response to bowel obstruction; thrombocytopenia and anaemia were attributed to the same process.

Plain supine and erect abdominal radiographs demonstrated a triple-bubble appearance with multiple dilated proximal small-bowel loops and air-fluid levels, in keeping with proximal small-bowel obstruction (Fig. 2). Abdominal ultrasonography was performed to exclude malrotation with midgut volvulus and ileocolic intussusception; both were excluded, and findings were considered consistent with intestinal atresia. A contrast enema was deferred in view of the unequivocal radiographic and clinical picture and the urgency of surgical intervention; however, intra-operative inspection of the colon was planned. Echocardiography and renal ultrasonography performed preoperatively did not demonstrate any associated cardiac or genitourinary anomalies.

Therapeutic intervention

The neonate was admitted to the surgical neonatal unit, where a nasogastric tube was placed for decompression, intravenous fluid resuscitation with isotonic crystalloid was commenced, and broad-spectrum antibiotic prophylaxis (ampicillin and gentamicin) was initiated. Vitamin K was administered. After approximately 12 hours, the dehydration had resolved, and biochemical parameters had improved sufficiently to permit anaesthesia. The infant was transferred to the operating theatre.

Under general endotracheal anaesthesia, a supra-umbilical transverse laparotomy was performed. Systematic exploration revealed six discrete atretic segments distributed along the distal jejunum and proximal-to-mid ileum, separated by short segments of grossly dilated proximal bowel and collapsed distal bowel. The anatomy corresponded to Grosfeld Type IV. The mesentery was intact, with no apple-peel deformity. The duodenum, stomach, colon, and Meckel's region were carefully inspected and were normal. The total length of small bowel from the ligament of Treitz to the ileocaecal valve was estimated

intra-operatively and judged to be adequate to permit single-stage resection. A bowel segment of approximately 30 cm encompassing all six atresiae was resected en bloc, and intestinal continuity was restored by a single-layer end-to-end interrupted anastomosis using 5 – 0 polydioxanone (Figs. 3 and 4). Patency was confirmed by gentle saline irrigation through the proximal limb and atraumatic passage of a soft catheter across the anastomosis. Haemostasis was secured, and the abdomen was closed in layers.

Postoperative course and follow-up

Postoperative management was standard. Bowel function returned on postoperative day 4, as evidenced by passage of meconium-stained then transitional stools. Trophic enteral feeding with expressed breast milk was introduced on day 5 and advanced gradually; full enteral feeding was achieved by postoperative day 10, at which point parenteral nutrition was discontinued. The patient was discharged on postoperative day 14 in good clinical condition, on full oral breastfeeding, and with documented appropriate weight gain.

At the most recent follow-up, 10 months postoperatively, the infant was thriving on age-appropriate complementary feeding. His weight was 9 kg, with appropriate length and head circumference. There were no clinical features of short-bowel syndrome, no episodes suggestive of adhesive obstruction, and no signs of anastomotic stricture. The mother reported no parental concerns regarding feeding or development. A structured surveillance plan, including periodic anthropometric assessment and clinical review, has been agreed with the family.

Discussion

Jejunoileal atresia is a well-recognised cause of congenital intestinal obstruction; however, Type IV with multiple atretic segments is comparatively uncommon, accounting for approximately one-third of multiple-atresia presentations and representing a smaller proportion of all JIA cases [4, 12]. The case reported here is noteworthy on two grounds: first, the presence of six discrete atretic segments (a number at the upper end of the range commonly described) and second, the absence of any associated anomaly. Most Type IV atresiae described in the literature are accompanied by extra-intestinal malformations (cardiac, genitourinary, or syndromic) or by additional gastrointestinal abnormalities such as malrotation, gastroschisis, or cystic fibrosis [2, 10]. Truly isolated Type IV disease, as in our patient, is therefore distinctly uncommon.

Pathogenesis

The vascular hypothesis advanced by Louw and Barnard remains the dominant explanation for jejunoileal atresia [7]. Multiple atresiae are thought to result either from a single but extensive ischaemic insult affecting a contiguous mesenteric territory, or from multifocal vascular events. Familial clustering

of multiple intestinal atresiae has been described and is associated with mutations in the TTC7A gene, often in conjunction with combined immunodeficiency; the sporadic occurrence and uncomplicated postoperative course in our patient argue against this entity, but long-term follow-up for immunological signs remains prudent [12].

Diagnostic considerations

Bilious vomiting in any neonate must be considered to indicate mechanical obstruction until proven otherwise [8]. The triad of bilious emesis, abdominal distension, and delayed meconium passage was present in our patient and is highly suggestive of distal small-bowel obstruction. Plain abdominal radiography remains the first-line imaging modality and was diagnostic in our case. Ultrasonography is operator-dependent but valuable for excluding midgut volvulus and intussusception, both of which constitute surgical emergencies with overlapping presentations [7]. A contrast enema may be helpful to differentiate small- from large-bowel obstruction and to identify an unused microcolon, which is characteristic of distal atresia; in the present case, the strong clinical suspicion and radiographic findings were considered sufficient to proceed to laparotomy without further imaging, and the integrity of the colon was confirmed intra-operatively. Echocardiography and renal ultrasonography are recommended in every confirmed case of intestinal atresia in order to identify associated anomalies [2, 10].

Surgical strategy: resection versus conservation

The optimal surgical approach to multiple atresiae is debated. Two broad strategies have been described in the literature [5, 6, 11]. The first is en-bloc resection of all atretic segments with a single primary anastomosis, which simplifies the operation, eliminates multiple anastomotic sites, and minimises the cumulative risk of anastomotic leak, but at the cost of additional bowel length. The second is bowel-sparing, with multiple anastomoses (with or without temporary enterostomy) to preserve as much length as possible, which is critical when the residual bowel is borderline. Govindarajan and Annamalai have explicitly framed this dilemma in terms of "resection versus conservation" and emphasised that the decision must be individualised on the basis of the total length of preserved healthy bowel, the distance between atretic segments, the calibre disparity, and the haemodynamic stability of the neonate [5].

In the present case, the six atresiae were closely grouped along a 30-cm segment, the residual small-bowel length was assessed intra-operatively as adequate, and the neonate was haemodynamically stable. These factors favoured single-stage en-bloc resection and primary anastomosis, which proved well tolerated. The postoperative outcome aligns with the findings of Goodluck and colleagues from northern Tanzania, who reported similar success with primary anastomosis in resource-limited settings, and with the systematic comparison by Eeftinck Schattenkerk et al., which suggests that staged enterostomy approaches are associated with a higher cumulative complication burden than primary anastomosis when the latter is feasible [10, 11].

Postoperative considerations and complications

Recognised postoperative complications include short-bowel syndrome, anastomotic leak, anastomotic stricture, functional dysmotility of the dilated proximal bowel, and adhesive obstruction [11]. Tapering or plication enteroplasty of a markedly dilated proximal limb is sometimes required to address calibre disparity and reduce the risk of functional obstruction; in our patient, the calibre mismatch was considered acceptable, and tapering was not performed. Long-term nutritional and developmental surveillance is essential, particularly when the resected length approaches or exceeds 30 cm in a neonate, given the risk of late short-bowel sequelae.

Strengths and limitations

The strengths of this report include adherence to the CARE 2013 reporting framework, intra-operative documentation of all six atretic segments, exclusion of associated anomalies through targeted imaging, and a structured 10-month follow-up demonstrating favourable growth and the absence of complications. The limitations include the retrospective nature of the report, the absence of formal genetic testing for TTC7A and related conditions, and a follow-up duration that, while informative for early outcomes, is insufficient to exclude late sequelae such as adhesive obstruction or growth faltering. Continued long-term follow-up is planned.

Conclusion

Isolated Type IV jejunoileal atresia is a rare congenital entity that requires a high index of clinical suspicion, prompt radiographic confirmation, and individualised surgical management. In appropriately selected neonates, en bloc resection of multiple closely grouped atretic segments with a single-stage primary end-to-end anastomosis is a safe and effective strategy that avoids the morbidity of stoma formation and multiple anastomoses. Long-term surveillance for nutritional, growth-related, and surgical complications remains essential, and clinicians should remain alert to the possibility of associated cardiac, genitourinary, syndromic, or genetic conditions.

Declarations

Acknowledgement

The authors would like to thank the medical and nursing staff involved in the care of this patient.

Ethical approval

In accordance with our institutional policy, single-patient anonymized case reports are exempt from formal Institutional Review Board approval, provided that written informed consent is obtained from the patient or legal guardian.

Consent

Written informed consent was obtained from the patient's parents for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal upon request.

Data availability

All data relevant to this case report are included within the article.

Conflicts of interest

The authors declare that they have no competing interests.

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Author contributions

All the authors equally contributed to the writing, drafting, writing, reviewing, conceptualizing, and investigation. All the authors approved the last version submitted to the journal.

References

1. Belkin-Gerson J, Pekarsky AR. Jejunoileal atresia. In: MSD Manual Professional Version [Internet]. 2025 [cited 2025 Jul 6]. Available from: https://www.msdmanuals.com/professional/pediatrics/congenital-gastrointestinal-anomalies/jejunoileal-atresia
2. Osuchukwu OO, Rentea RM. Ileal Atresia. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 [updated 2023; cited 2025 Jul 6]. Available from: https://www.ncbi.nlm.nih.gov/books/NBK557400/
3. Adams SD, Stanton MP. Malrotation and intestinal atresias. Early Hum Dev. 2014;90(12):921–5. doi:10.1016/j.earlhumdev.2014.09.017
4. Grosfeld JL, Ballantine TV, Shoemaker R. Operative management of intestinal atresia and stenosis based on pathologic findings. J Pediatr Surg. 1979;14(3):368–75. doi:10.1016/s0022-3468(79)80502-3
5. Govindarajan KK, Annamalai M. Multiple small bowel atresia: resection or conservation? J Coll Physicians Surg Pak. 2021;31(6):740–2. doi:10.29271/jcpsp.2021.06.740
6. Oh C. Jejunoileal atresia: a contemporary review. Adv Pediatr Surg. 2023;29(2):89–98. doi:10.13029/aps.2023.29.2.89
7. Shalkow J. Small intestinal atresia and stenosis. Medscape [Internet]. 2023 Sep 28 [cited 2025 Jul 6]. Available from: https://emedicine.medscape.com/article/939258-overview

8. Aihole JS. A rare case of jejunal atresia. Int J Surg Case Rep. 2022;90:106714. doi:10.1016/j.ijscr.2021.106714
9. National Organization for Rare Disorders. Jejunal atresia [Internet]. NORD; 2020 Jun 18 [cited 2025 Jul 6]. Available from: https://rarediseases.org/rare-diseases/jejunal-atresia/
10. Goodluck G, Gnanamuttupulle M, Sadiq A, Msuya D, Chugulu S, Lodhia J. Jejunoileal atresia in a newborn: our experience from northern Tanzania and literature review. Clin Med Insights Pediatr. 2022;16:11795565221075313. doi:10.1177/11795565221075313
11. Eeftinck Schattenkerk LD, Musters GD, de Jonge WJ, et al. Treatment of jejunoileal atresia by primary anastomosis or enterostomy: double the operations, double the risk of complications. J Pediatr Surg. 2022;57(9):49–54. doi:10.1016/j.jpedsurg.2021.07.021
12. Aguayo P, Ostlie DJ. Duodenal and intestinal atresia and stenosis. In: Holcomb GW, Murphy JP, Ostlie DJ, editors. Ashcraft's Pediatric Surgery. 5th ed. Philadelphia: W.B. Saunders; 2010. p. 400–15. doi:10.1016/B978-1-4160-6127-4.00031-8

Figures



Figure 1

Clinical photograph on admission demonstrating bilious vomiting following the breastfeeding process.

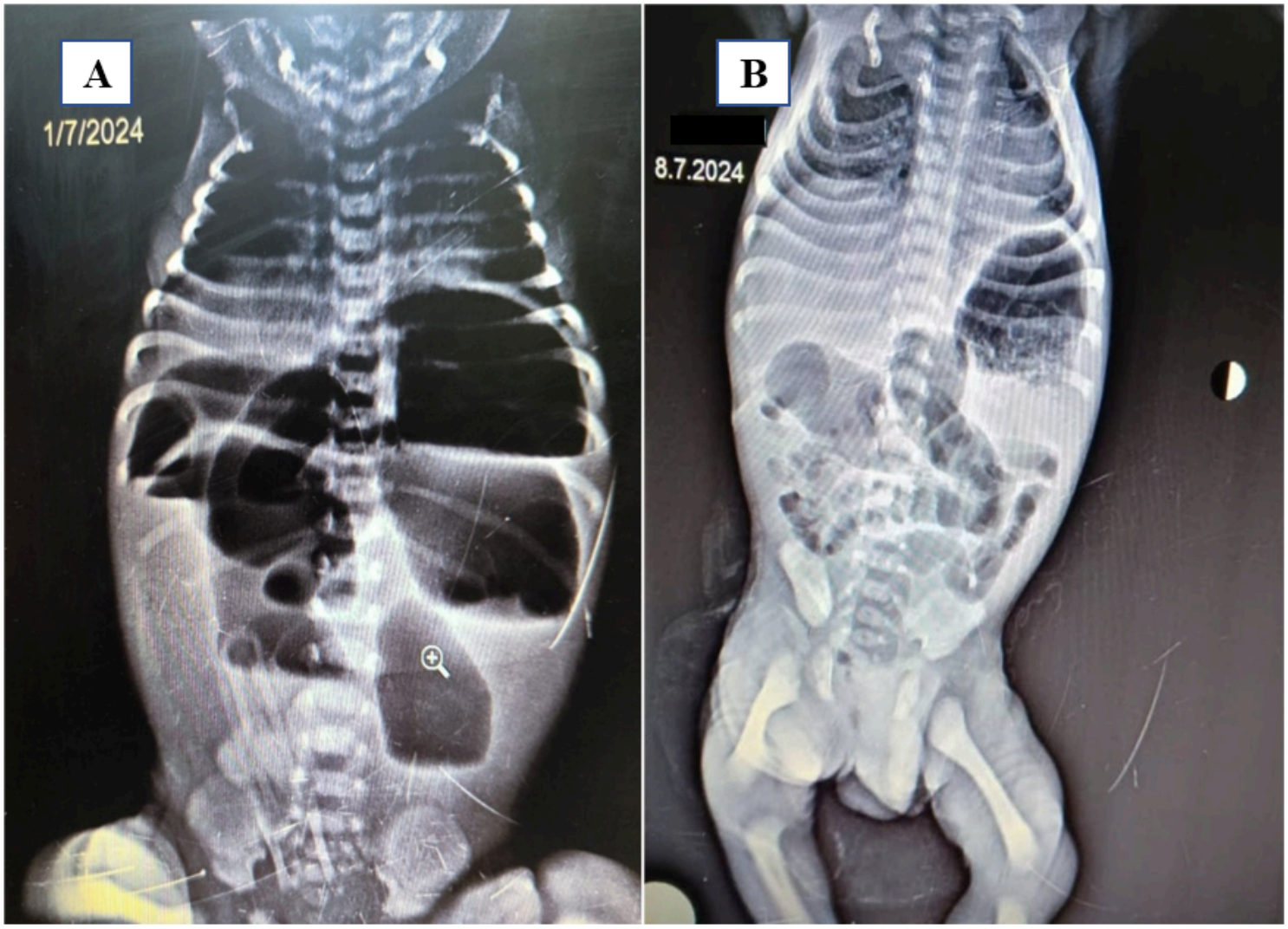


Figure 2

Abdominal X-ray reveals significant signs indicative of multiple obstructions in the jejunoileal.

(A) a triple bubble sign suggestive of atresiae.

(B) multiple dilated jejunoileal loops with multiple air fluid levels suggestive of obstruction.

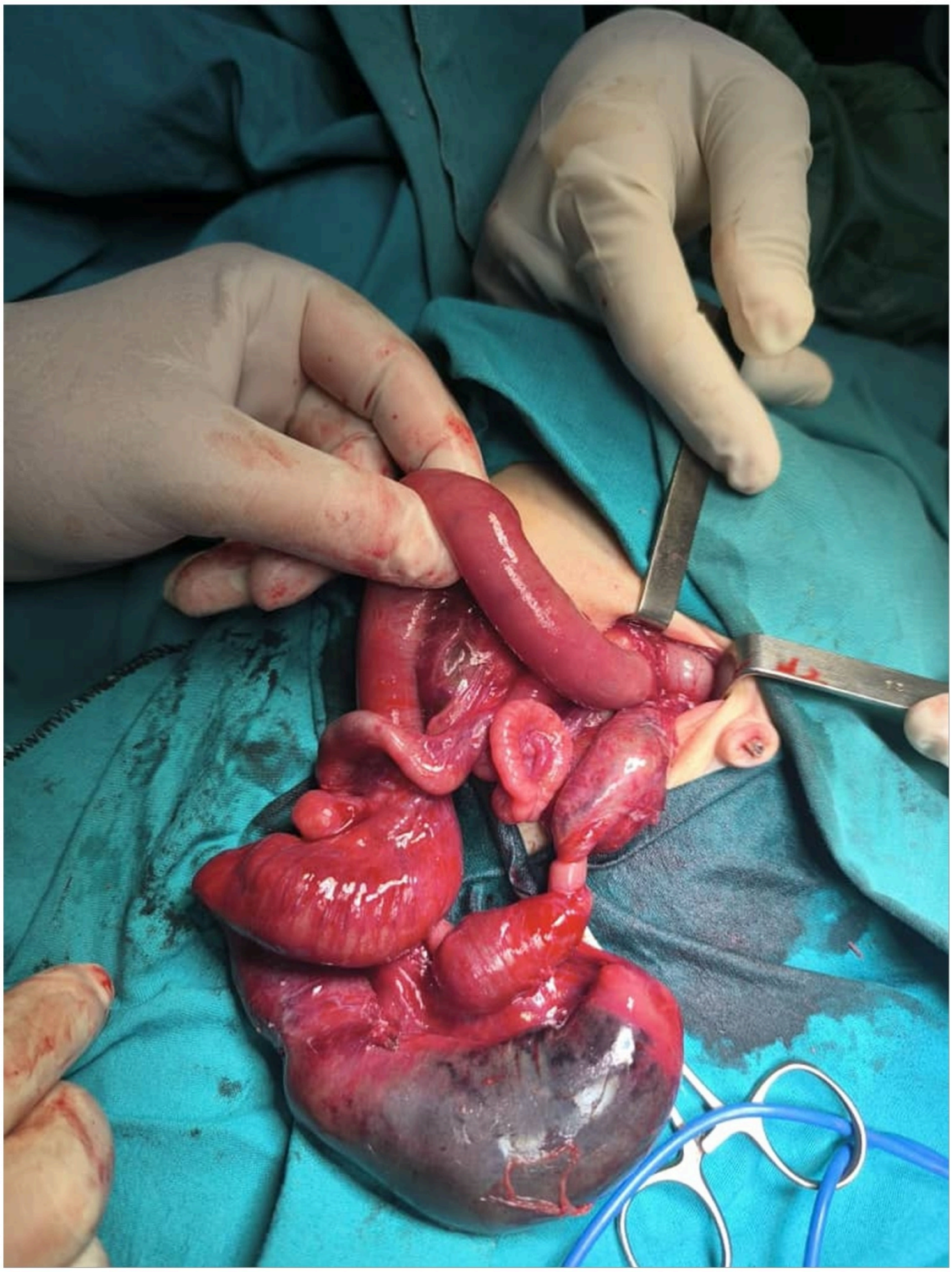


Figure 3

Intra-operative photograph illustrating the six atretic jejunoileal segments along a closely grouped portion of small bowel.

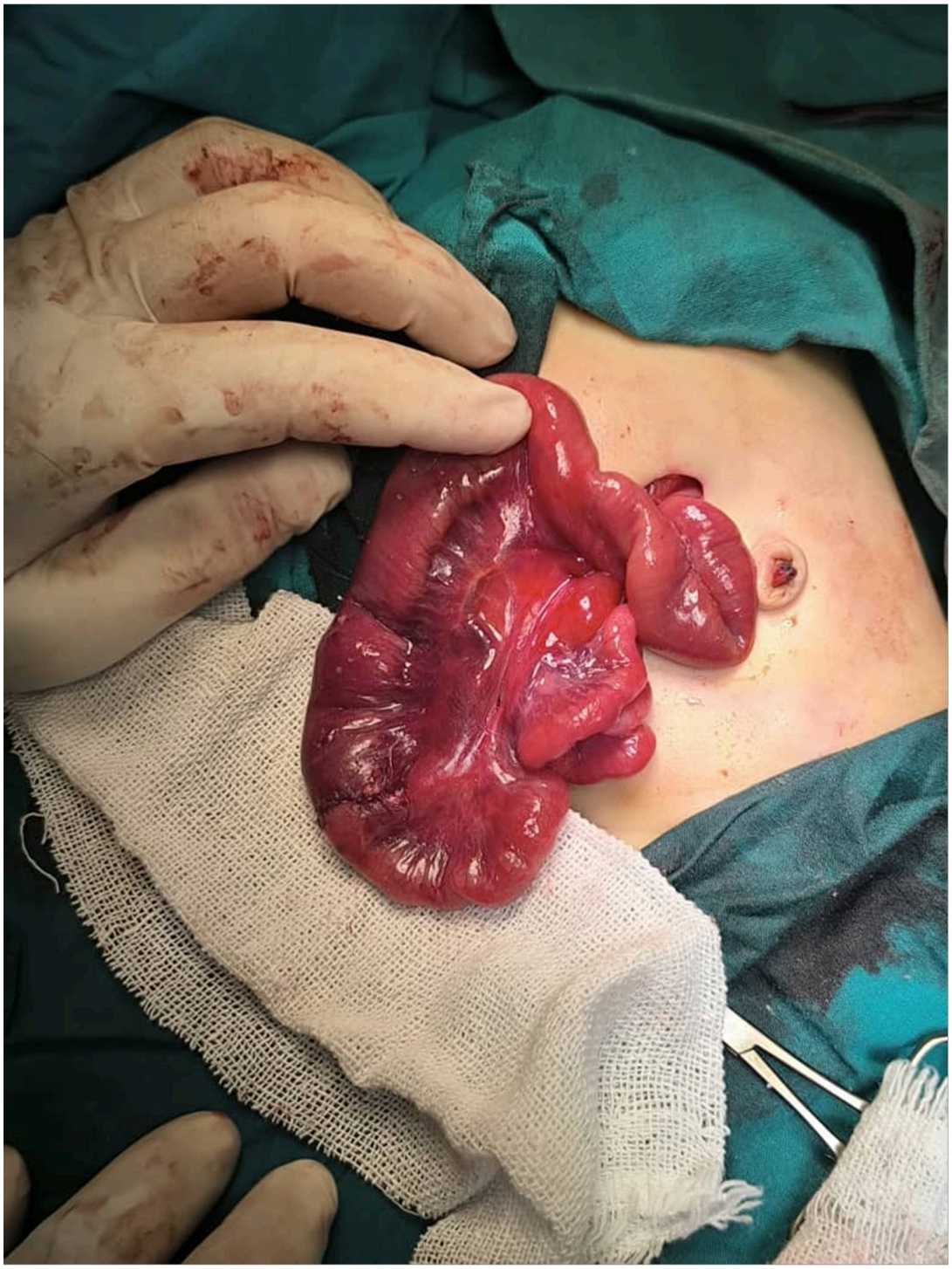


Figure 4

Intra-operative photograph after en-bloc resection (approximately 30 cm) and single-layer end-to-end primary anastomosis.

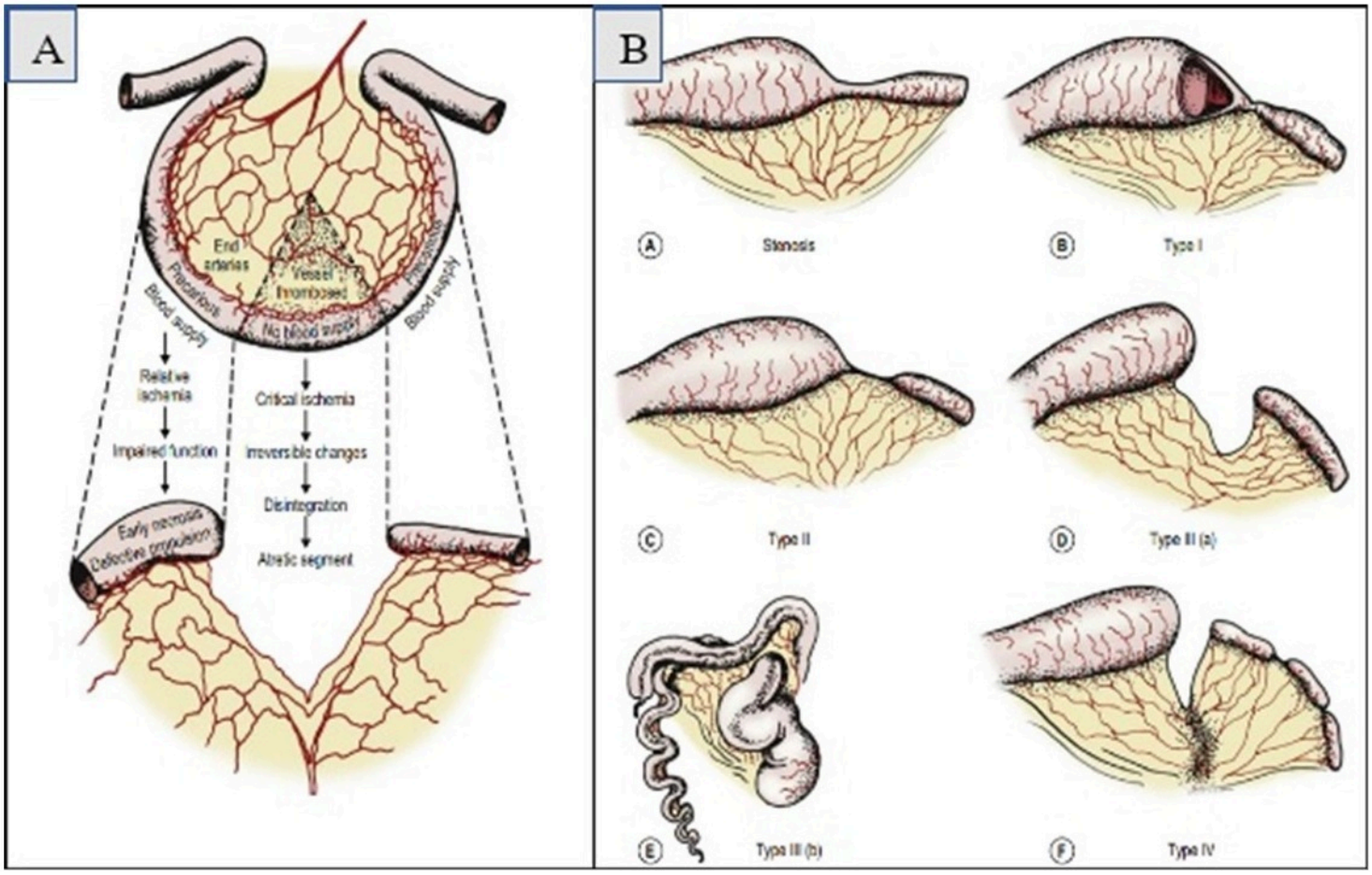


Figure 5

The Figures were taken from " Holcomb and Ashcraft's Pediatric Surgery, 7th Edition " [11]

(A) the hypothesized mechanism of vascular compromise leading to the subsequent development of JIA

(B) The classification system for jejunoileal atresia and stenosis.