CLINICAL SURGERY JOURNAL

Clinical Surgery Journal

ISSN: 2767-0023

Congenital Neonatal Proximal Jejunal Isolated Segmental Dilatation

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Article Type: Case Report

Compiled Date: November 03, 2026

Volume: 7

Issue: 2

Journal Name: Clinical Surgery Journal

Journal Short Name: Clin Surg J

Publisher: Infact Publications LLC

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Keywords: Acute abdomen; Neonatal; Prenatal; Congenital; Intestinal obstruction; Jejunal; Segmental dilatation; Periumbilical laparotomy

Cite this article: Govani DR, Singh H, Swamy KB, Midha PK, Govani ND, Panchasara NG, et al. Congenital Neonatal Proximal Jejunal Isolated Segmental Dilatation. Clin Surg J. 2026;7(2):1–3.

Abstract

We present a 3600 g male term neonate born by spontaneous vaginal delivery who had polyhydramnios and a dilated bowel loop suggestive of intestinal obstruction on prenatal scans. The baby presented with neonatal intestinal obstruction at birth, with bilious vomiting and no abdominal distension, mimicking midgut volvulus. A plain abdominal radiograph demonstrated an unexplained massively dilated bowel loop with gas in the stomach proximally and gas-filled bowel loops distally. Abdominal ultrasound and an upper gastrointestinal water-soluble contrast study identified a massive segmental dilatation of the proximal jejunum causing functional partial upper small bowel obstruction. Emergency exploratory laparotomy after resuscitation confirmed an isolated proximal jejunal massive segmental dilatation, and resection of this dilated segment with end-to-end anastomosis was performed. Post-operative recovery was rapid, and at long-term follow-up the patient remains asymptomatic, thriving well, with normal abdominal examination and ultrasound scan at 20 years of age.

Introduction

Congenital Segmental Intestinal Dilatation (CSID) is a rare anomaly that can involve any part of the intestine, though the ileum is most commonly affected. Proximal jejunal segmental dilatation is extremely rare [1,2]. Diagnosis may occur prenatally using ultrasound, in the neonatal period or early infancy when symptoms of partial or total obstruction arise, or later in childhood and adulthood. A case of prenatally diagnosed intestinal obstruction caused by isolated segmental dilatation of the proximal jejunum is exceptionally rare, particularly when presenting in the immediate neonatal period as acute intestinal obstruction.

Case Presentation

A one day old male neonate, born at term weighing 3600 g, was delivered spontaneously following antenatal findings of polyhydramnios and a dilated bowel loop suggestive of intestinal obstruction. Initial evaluation was performed at a district hospital before transfer to our Neonatal Intensive Care Unit (NICU). The infant developed bilious vomiting immediately after birth. He was clinically stable, with upper abdominal distension that was soft and non tender, and no palpable mass. Blood counts and biochemical profiles were normal. Post natal ultrasound confirmed a massively dilated proximal jejunal segment with no associated anomalies. Plain abdominal radiograph revealed a massively dilated bowel loop in the upper abdomen, with gas visible both proximally in the and distally in the pelvis (Figure A).

Differential diagnoses included acute intestinal obstruction, volvulus, Hirschsprung's disease, and midgut malrotation. To exclude midgut volvulus, a potentially lethal condition, a water soluble contrast study was performed via nasogastric tube. An upper gastrointestinal contrast study confirmed a sharply demarcated dilated segment of the proximal jejunum just beyond the duodenojejunal flexure. This confirmed an adynamic, massively dilated, isolated proximal jejunal loop without mechanical obstruction, with abundant bowel gas distally in the abdomen and pelvis (Figure B).

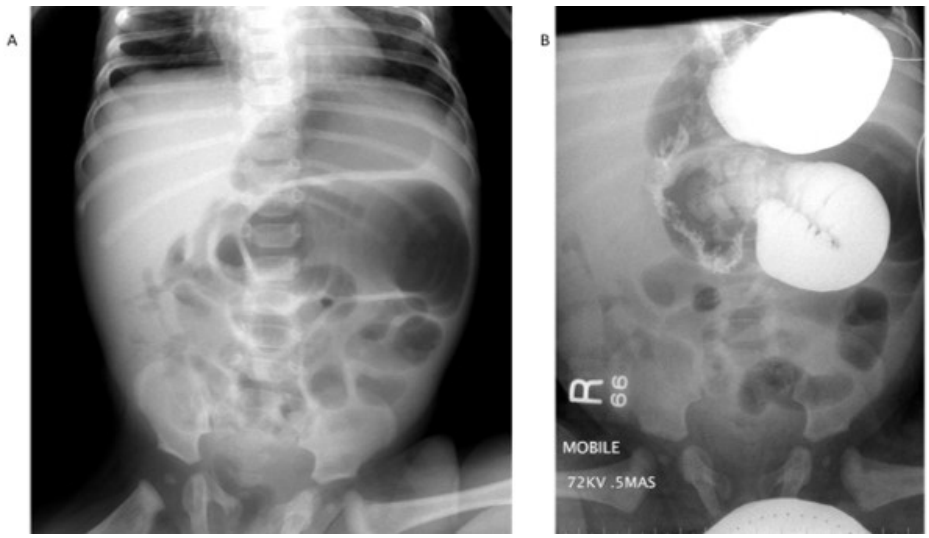


Figure: Radiographic and contrast imaging of congenital proximal jejunal segmental dilatation

A) Plain abdominal radiograph demonstrating a massively dilated bowel loop in the upper abdomen, with gas visible in the stomach proximally and in distal bowel loops, suggestive of partial intestinal obstruction.

B) Upper gastrointestinal contrast study revealing a sharply demarcated, dilated segment of the proximal jejunum located just beyond the duodenojejunal flexure, consistent with isolated segmental dilatation.

The infant was resuscitated with intravenous fluids and nasogastric suction. Emergency exploratory laparotomy via a minimally invasive periumbilical incision revealed a hugely dilated isolated proximal jejunal segment distal to the duodenojejunal flexure. No obstructing lesion was present, and bowel contents could be moved freely.

Resection of the dilated segment followed by end to end jejunojejunostomy was performed uneventfully. Post operative recovery was smooth. Breastfeeding commenced on day 5, and discharge occurred on day 7. At 20 year follow-up, the patient remains well and asymptomatic.

Discussion

Congenital segmental jejunal dilatation presenting as neonatal intestinal obstruction is rare. Congenital Segmental Intestinal Dilatation is characterized by a localized, sharply demarcated dilated segment of intestine without intrinsic or extrinsic obstruction, abnormal innervation, or transition zone.

In neonates, CSID may mimic midgut volvulus, Hirschsprung's disease, or functional obstruction [3]. Jejunal involvement is uncommon, and its etiology remains unclear despite several proposed mechanisms [4]. These include intrauterine vascular accidents [5], associations with multiple intestinal atresias [6], and external compression of the fetal bowel [7].

Swenson and Rathauser (1959) [8] established diagnostic criteria:

- Abrupt transition between dilated and normal bowel.
- No intrinsic or extrinsic barrier distal to the dilatation.
- Limited bowel dilatation with a 3- to 4-fold increase in size.
- Clinical presentation of occlusion or sub-occlusion.
- Complete recovery after resection.
- Normal neuronal plexus.

A nationwide retrospective cohort study reported 28 cases of CSID [9,10]. The ileum was affected in 14 cases, the colon in 10, and the jejunum rarely. Surgical resection with end-to-end anastomosis was the treatment of choice in nearly all cases, with good prognosis.

Histological findings vary, including ectopic tissue and abnormal muscle layers. Rarely, immature ganglion cells have been reported in isolated jejunal dilatation [11]. Segmental stenosis with neonatal perforation has also been described [12]. Surgical resection remains the definitive treatment, with excellent outcomes when no associated anomalies are present.

Conclusion

This case highlights an unusual and rare presentation of isolated proximal jejunal segmental dilatation diagnosed prenatally and treated successfully in the immediate neonatal period. Resection and end to end anastomosis resulted in an excellent long term

prognosis.

Acknowledgement

We are grateful to Mr. Clive W. Hardy for proofreading our article and making extensive language corrections. We are grateful to Miss Indre Zaparackaite, Consultant Pediatric Surgeon at the Evelina Children's Hospital, London; Mr. Shailinder Singh, Consultant Pediatric Surgeon at the University of Nottingham NHS Foundation Trust, Nottingham, UK; Prof. C K Sinha, Consultant Pediatric Surgeon at St George's Hospital, London; and Dr. Debashish Bhattacharya, Chief Pediatric Surgeon at Sultan Qaboos University Teaching Hospital, Salalah, Oman, who were involved in drafting, revising, and editing the manuscript critically for important intellectual content.

Conflict of Interests

The authors declare no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.
Informed consent was obtained for this publication.

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