Case Report

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KEYWORDS
Segmental dilatation, Jejunum, Giant, Malrotation

ABSTRACT
Background: Giant segmental dilatation of jejunum (SDJ) is rare in small infants where it often presents with intestinal obstruction. Occasionally, the giant SDJ may twist resulting in volvulus.

Case Presentation: A 1-month-old female infant presented with intestinal obstruction. At surgery, isolated giant segmental dilatation of the jejunum causing volvulus of the bowel was found. The gut was viable. The SDJ was excised and jejuno-jejunal anastomosis was performed. Ladd’s procedure was also performed for the associated malrotation. Postoperative recovery remained uneventful.

Conclusion: Giant SDJ is a rare entity, rarer still is its presentation with volvulus secondary to its twist. Early diagnosis and prompt management result in a good outcome.

INTRODUCTION
Congenital segmental dilatation (CSD) of the intestine is rare and first described by Swenson and Rathausser in 1959. [1] Over 150 cases have been reported so far. [2] The dilated segment has impaired motility irrespective of having normal innervation and ganglion cells, though it usually has abnormalities in the muscular layer. [3]

Jejunum is rarely involved as the ileum and colon are the most typical site of occurrence. In older children, it presents with intractable constipation while in small infants and neonates, the presentation is with intestinal obstruction. [4] Herein, we present a case of giant segmental dilatation of jejunum (SDJ) that presented with intestinal obstruction secondary to its volvulus.

CASE REPORT
A 1-month-old female infant presented with absolute constipation and bilious vomiting for 2 days. She was a full-term baby delivered through C-Section. The patient initially had non-bilious vomiting associated with milk intake, which later became bilious in the last 18 hrs in multiple episodes. The patient was dehydrated on examinations with sunken eyes and dry coarse skin. The abdomen was tense and distended at the epigastric region. Bowel sounds were sluggish on auscultation. On per-rectal examination, the rectum was found collapsed.

Laboratory investigations showed anemia (Hb 6.7g/dL), and leukocytosis (19.6*10^3/uL). Serum sodium was 132 (135-145) mol/L and potassium was 2.9(3.4- 5.5) mmol/L. X-ray abdomen showed a distended, enlarged gastric bubble. There are some air-fluid levels along with the ground glass appearance of the abdomen (Fig. 1A). Ultrasound abdomen showed a cystic lesion approximately 5.6*4.5cm in the epigastric region. Due to solid suspicion of volvulus, we didn’t perform a contrast study.

After taking informed consent and resuscitations, the patient was shifted to the operation theater and had an open exploration with the right-sided supraumbilical transverse incision. There was isolated jejunal dilatation approximately 5*5cm in size, 10cm from the duodenjejunal junction, having prominent serosal vessels around which the gut was twisted (Fig. 1B). Cecum was mobile and DJ was on the right side of the spine with multiple Ladd bands. The volvulus was not related to malrotation but rather secondary to segmental dilatation. The isolated segment of the jejunum on resection showed a single lumen with an abrupt transition to normal caliber bowel on either side. The SDJ was resected after de-twisting of the gut, and jejuno-jejunal anastomosis along with Ladd’s...
procedure was performed. The patient was shifted to the surgical intensive care unit for meticulous monitoring. Post-operative recovery was smooth and uneventful. The patient was discharged on the 6th postoperative day. Histopathology showed thinned-out bowel mucosa with ulcerations and underlying dense inflammation (Fig. 1C & 1D). The patient is doing fine on follow-up.

![Image of X-ray, operative picture, and histopathology](image)

**DISCUSSION**

Congenital segmental dilatation of the gut is a rare condition, and the literature has documented more than 150 cases from all around the globe. Still, none of them provide any clue regarding its etiology. [5]

Segmental dilatation can affect any part from the duodenum to the distal colon. The ileum is the most often affected region (50–61.1% of cases), followed by the jejunum (9.7–10.7%) and, less frequently, the duodenum (2.8–3.6%). [6] Segmental dilatation of the jejunum was first reported by Rossi and Giacomoni in 1973. [7]

It can present early in the neonatal period with obstruction and mimic volvulus or Hirschsprung disease and late with chronic constipation, anemia, malabsorption, or signs of intermittent intestinal obstruction. [4,8] Recurrent abdominal pain and features of obstruction are uncommon presentations. Some of them remain symptoms free till late adulthood. In our case, the patient had an acute presentation with absolute constipation and bilious vomiting for 2 days.

Ratcliffe et al. [9] reviewed 36 cases, 50% of the patients had gastrointestinal tract anomalies, including Meckel's diverticulum in seven instances, omphalocele in six cases, and malrotation of the gut in six cases. In the index case, only malrotation was found as an associated anomaly.

The diagnostic criteria for this uncommon entity were developed in 1959 by Swenson and Rathauer. [1] Criteria included (i) a limited bowel dilatation with a 3–4 folds increase in size, (ii) an abrupt change from dilated to normal bowel, (iii) no intrinsic or extrinsic barrier distal to the dilatation, (iv) a clinical picture of intestinal occlusion or sub occlusion, (v) a normal neuronal plexus, and (vi) full recovery following resection of the affected segment. In our case, bowel dilatation was 3-4 sizes of normal gut, an abrupt change from dilated to normal bowel, and no intrinsic or extrinsic barrier distal to the dilatation. Despite these criteria, variability in size and shape has been reported in the literature.[9] Mirza et al. have classified SDJ into five categories based on morphology i.e., Forme Fruste or pseudo-dilatation, Fusiform, saccular, tubular, and complex. [10] The index case belongs to the Saccular variety.

Some authors have also documented the presence of heterotopic tissues in the dilated segment, including striated muscle, esophageal, gastric, lung, and pancreatic tissues. [11] There have also been reports of a lack of muscularis propria entirely or partially in the affected area. [12] Cheng et al. carried out histochemical and immunohistochemical analyses, which revealed localized vacuolization of the intestinal smooth muscle in their case and suggested myopathy as the cause of dilatation. [13] Localized myopathy that contributes to segmental dilatation may also have interstitial cells of Cajal as a contributing cause. Ganglionic dysplasia is also suggested by some authors as a cause of congenital segmental dilatation.[14] In our index case, histopathology showed thinned-out mucosa with tall columnar mucinous type intestinal lining and underlying muscle coat. There is moderate mixed inflammation. No evidence of granuloma, dysplasia, and malignancy was seen.

Congenital segmental dilatation is an uncommon condition with a variety of morphological variants and the only effective treatment is total surgical excision with end-to-end anastomosis.[10] In critically sick patients, stoma can be made with resection of the dilated segment.[15] Tailored surgical strategies must be applied in more complex and unique cases.

In conclusion, SDJ is a rarer entity and rarer still is its presentation with the twist of the bowel secondary to it. Early diagnosis and prompt management yield good outcomes.

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