

## Case Report

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## Congenital segmental dilatation of intestine with different morphology: A case report

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## KEYWORDS

Neonatal intestinal obstruction,  
Anorectal malformation,  
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## ABSTRACT

**Background:** Congenital segmental dilatation of intestine is a rare entity characterized by a localized dilated segment of the intestine. It manifests as neonatal intestinal obstruction. Occurrence with another surgical anomaly may masquerade it and complicate the treatment course.

**Case Presentation:** A 1-day-old male neonate with anorectal malformation underwent colostomy as first stage of the treatment. The stoma did not function postoperatively. At repeat surgery, congenital segmental dilatation of the ileum, with slightly different morphology, was found. Resection of the lesion and ileostomy were formed. Postoperative course remained uneventful.

**Conclusion:** Congenital segmental dilatation of intestine may be missed at initial surgery. Complete surgical resection is curative.

## INTRODUCTION

Congenital segmental dilatation of intestine is rare with over 150 cases reported so far.[1] The normal intestine abruptly transits to a dilated segment of variable length and diameter (3-4 times) without any evidence of mechanical obstruction.[2] The dilated segment is hypomotile but with normal neuronal connections and ganglion cells. Often, it is a solitary lesion, but multiple sites may be involved.[3] Ileum is the most common site followed by colon, jejunum, and duodenum.[4] The extent may range from a small lesion to as much as near-total colon.[2]

Swenson et al. proposed diagnostic criteria of congenital segmental dilatation of the intestine; and most of the reported cases followed it, except a few.[5] Herein, we report a morphologically different case of congenital segmental dilatation of ileum in a patient with anorectal malformation.

## CASE REPORT

A 1-day-old male neonate, weighing 2.5kg, presented with failure to pass meconium since birth. The baby was born at term by spontaneous vaginal delivery. Antenatal follow-up and perinatal period were uneventful. On examination, the baby had abdominal distension and an imperforated anus. X-ray abdomen

(prone cross-table lateral, and erect) showed a high variety of anorectal malformation (Fig. 1A, 1B). Ultrasound of the abdomen revealed normal kidneys. Echocardiography showed a small PDA. The laboratory tests were within normal ranges.

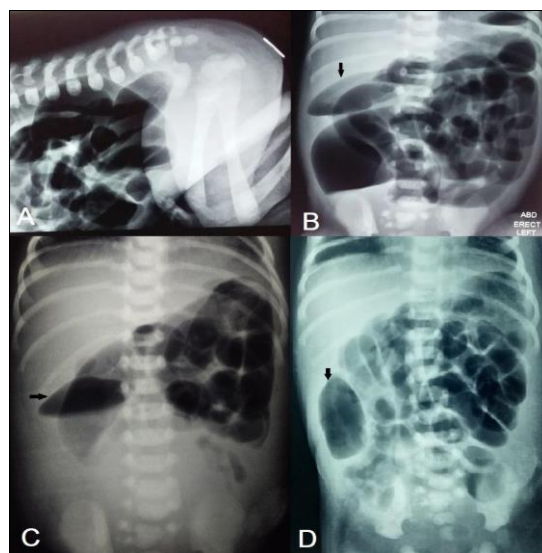


Figure 1: A) Cross table lateral film showing high anorectal malformation. B) Preoperative erect radiograph showing a fusiform area (arrow). C) Postoperative erect radiograph showing the same finding (arrow). D) Postoperative supine radiograph showing the same finding (arrow).

After optimization, the patient underwent sigmoid loop colostomy over a skin bridge as first stage surgery in the management of high variety anorectal malformation. The next day, the patient passed negligible meconium through the stoma. The abdomen further distended and stoma was non-functional.

The repeat X-ray abdomen showed dilated small bowel loops. A fusiform area of bowel gas was noted in the right hemiabdomen that was also present on the initial radiographs. Laboratory tests showed a rising C-reactive protein and declining thrombocytes. The next day, repeated x-rays had the same findings; thus, the parents were counseled for reoperation. A segmental dilated portion of mid-ileum was found at exploratory laparotomy with proximal dilated loops and distal comparatively collapsed bowel (Fig. 2). The abrupt transition from normal bowel to segmental dilatation was absent. The dilated segment was resected, and an ileostomy formed—anastomosis avoided as the baby was getting septic. Serial biopsies were taken from the ileostomy site and colon that excluded Hirschsprung's disease.



Figure 2: Showing Segmental dilatation of ileum (arrow).

The postoperative recovery remained uneventful. The baby passed adequate stool from ileostomy and was discharged in good condition on ad libitum. The patient is on our follow-up for definitive surgery. We have prioritized ileostomy reversal first, followed by anorectoplasty and colostomy reversal.

## DISCUSSION

Congenital segmental dilatation of intestine encounters mostly in neonates (>60%) with a male preponderance.[4] Finding merely case reports and small case series in the literature vindicates its scarcity.[6] Sakaguchi et al. identified 28 cases in a nationwide survey of Japan, over a decade period.[4] We also published a case report in 2012 and a series of 3 cases in 2016.[3,6]

Two patterns of presentation exist: early with neonatal intestinal obstruction; and late with chronic constipation and related sequelae such as failure to

thrive, malnutrition, malabsorption, anemia, etc.[2,4,7,8] Ileal segmental dilatation usually presents early whereas colonic segmental dilatation appears to escape the early presentation, though not a rule.[2,6,9]

Associated anomalies may alter the presentation as in the index case; we initially missed the segmental dilatation because the colostomy was formed with a small incision; later we identified it on exploratory laparotomy for persistent intestinal obstruction. We have reported a similar case of imperforate anus with congenital pouch colon where the segmental dilatation was identified on repeat surgery for early postoperative intestinal obstruction.[6]

Antenatal and preoperative diagnoses are seldom, with most cases diagnosed at surgery.[4,7] Abdominal X-ray, ultrasound, CT scan, and GIT contrast study may help the diagnosis preoperatively; though needs a high index of suspicion. It can mimic other surgical conditions on radiological investigations: pneumoperitoneum on an abdominal radiograph,[6] and duplication cyst on contrast imaging are few examples.[10] In the index case, we initially ignored the discrete fusiform gas shadow on the abdominal radiographs and noticed when the finding persisted on subsequent radiographs.

Table 1: Morphological spectrum of congenital segmental dilatation of intestine identified on literature search

Type	Title	Description
I	Forme Fruste or pseudo-dilatation	Smaller lesions without abrupt transition to the segmental dilatation.(5)
II	Fusiform	Fusiform shape with more gradual transition to the segmental dilatation.(6)
III	Saccular	Rounded or sac-like bigger lesions with abrupt transition.(11)
IV	Tubular	Longer lesions with any type of transition.(7,9,12)
V	Complex	Multiple segmental dilatations(3) or associated with congenital pouch colon.(13)

On literature search, we have identified and classified the varied morphology of the congenital segmental dilatation of the intestine (Table 1). The diagnostic criteria laid by Swensen et al. appear exclusive, but we believe the anomaly has a developmental spectrum, and reported variations in morphology are common.[5,9] The reported cases differ in site, size, shape, and histology.

Adhikari et al. reported a case of a small congenital segmental dilatation of the jejunum which lacked an abrupt transition between the normal bowel and the

segmental dilatation.[5] They initially left it unresected as the mechanical obstruction was absent; on persistent postoperative intestinal obstruction, the lesion was resected at repeat surgery. Such lesions may represent a milder morphological variation (forme fruste) in the spectrum. The index case also appears to be a forme fruste of congenital segmental dilatation of intestine.

Complete surgical resection and anastomosis is curative; however at certain locations (such as the duodenum), tapering is recommended.[4,7] Occasionally a temporary stoma is needed in cases who are not fit or in sepsis.[9] Similarly in the index case, as the baby was in sepsis, we avoided anastomosis and a stoma was formed.

To conclude, congenital segmental dilatation of the intestine is a rare entity with morphological varia-

tions. Rarer still is its forme fruste variant. Occasionally these small lesions are missed on initial surgery thus complicating the treatment course. Complete surgical resection is the treatment of choice in most location.

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