

CONGENITAL SEGMENTAL DILATATION OF COLON

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Segmented dilatation of the colon belongs to the group of poorly understood dysmotility disorders of colon related to Hirschsprung's disease but without abnormality of ganglion cells. Of the fourteen previously reported cases of segmental dilatation of colon in the English literature, only two had presented in the neonatal age(1,2). We report a similar case and highlight the features common to this entity and congenital short colon seen in association with imperforate anus.

Case Report

A full term, 3800 g male was born by normal vaginal delivery to a 24-year-old primigravida following an uncomplicated pregnancy. The neonate presented at sixty hours of age with a history of abdominal distension, respiratory distress and cyanosis

for one day. He had vomited non-bilious material once and had passed small amount of meconium once. There was history of severe birth asphyxia. Initial examination revealed obvious dysmorphism, frontal bossing, flat facial profile, antimongoloid slant, bilateral subconjunctival hemorrhages, short neck and chest with pectus carinatum. The abdomen was grossly distended, tympanitic and no mass could be palpated. Rectal examination was essentially normal, finger stall was stained with meconium.

Initial investigations revealed thrombocytopenia (platelet count 15000/mm³), deranged bleeding profile (PT 227 13", PTTK 1' 15"/38"), raised blood urea (225 mg/dl) and serum creatinine (3.8 mg/dl). Serum electrolytes were normal. An erect abdominal radiography revealed a large air shadow occupying most of the abdomen, displacing the stomach and the small bowel (*Fig. 1*). X-ray also revealed multiple vertebral anomalies of thoracolumbo-sacral region; twelfth rib on the right side was missing. Barium enema demonstrated normal calibre rectum and sigmoid colon which abruptly opened into a large cystic dilatation of colon (*Fig. 2*). No definite diagnosis could be made and differential diagnosis of Hirschsprung's disease, duplication cyst of colon and segmental dilatation of colon was kept in mind.

After initial resuscitation the child was taken for laparotomy which revealed the loops of small bowel pushed into the right upper abdominal quadrant. The ileum opened into a normal cecum, from which a normal appendix arose. Then there was an abrupt transition to an enormously distended segment of colon which represented the ascending, transverse and descending colon. Distally, there was another abrupt transition to sigmoid colon which was of

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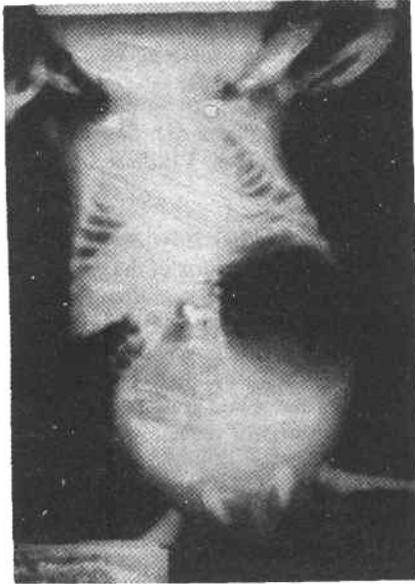


Fig. 1. Abdominal radiograph revealing a large air shadow occupying most of the abdomen and displacing the stomach and the small bowel.

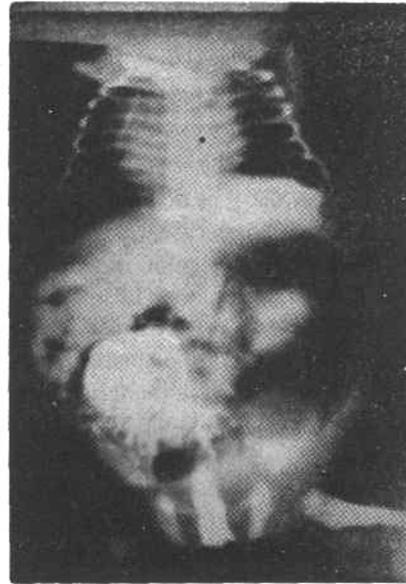


Fig. 2. Barium enema demonstrating massively dilated segment of colon. Note normal configuration of rectal ampulla and lower sigmoid.

normal calibre. *Teniae coli* were not demonstrable in the dilated segment, but were normal in both proximal and distal colon. The other abnormalities noted were Meckel's diverticulum and hydrops gall bladder. A loop ileostomy was done just proximal to Meckel's diverticulum and a biopsy was taken from the dilated colon. Resection of the dilated segment was not attempted keeping in view the poor general condition of the child.

Microscopic examination of the colon revealed presence of ganglion cells. The muscle layers were not hypertrophied although the arrangement of muscle was rather haphazard. The child continued to have episodes of apnea and cyanosis. A central cause, probably an intraventricular

hemorrhage was suspected for the same. The child succumbed to *Acinetobacter* septicemia and disseminated intravascular coagulation 7 days after surgery.

Discussion

Segmental dilatation of the colon was first described by Swenson and Rathauer in 1959(3). A review of English literature revealed eleven more cases since then(1,2,4-12).

Of the cases reported so far, age of presentation ranged from birth to 22 years. The clinical picture is similar to that of Hirschsprung's disease. The salient feature is the presence of a single, well defined segment of dilated colon without any evidence of intrinsic obstruction or of deficient

innervation and with more or less abrupt transition to normal bowel both proximally and distally. The side of dilated colon is variable, usually the left colon is involved. Definitive management consists of segmental resection with end-to-end anastomosis.

Based on the fact that symptoms in all the cases started very early in life, it is presumed that segmental dilatation is a congenital malformation(1,2). Our case lends further support to this assumption. The exact etiology of the condition remains unclear, although an intrauterine vascular catastrophe has been implicated(1,5).

When we compare the gross intraoperative features and histologic features to that of 'Congenital short colon associated with imperforate anus' (Zachary-Morgan Syndrome), there is a striking similarity between the dilated segment of two entities. The shortened colon (pouch colon) also lacks in *teniae coli*, but has histopathologically normal colonic wall including normal colonic innervation(13). The vascular pattern in cases of pouch colon is also irregular distorted and tortuous and is usually supplied by enlarged marginal colonic artery. This is similar to what is seen in some cases of segmental dilatation of colon(1,3).

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