Congenital segmental dilatation of the colon

Maher M. Al-Zaiem, Abdulhadi F. Al-Garni, Asim A. Asghar, Feras M. Al-Zaiem and Hatim AL-Omari

Congenital segmental dilatation of the colon is a rare cause of intestinal obstruction in neonates. We report a case of congenital segmental dilatation of the colon and highlight the clinical, radiological, and histopathological features of this entity. Proper surgical treatment was initiated on the basis of preoperative radiological findings. *Ann Pediatr Surg* 11:46–48 © 2015 Annals of Pediatric Surgery.

Annals of Pediatric Surgery 2015, 11:46–48

Keywords: congenital segmental dilatation of the colon, segmental dilation of intestines, neonatal intestinal obstruction

Pediatric Surgery Department, Maternity and Children Hospital, Mecca, Kingdom of Saudi Arabia

Correspondence to Maher M. Al-Zaiem, CU, ABPS, Pediatric Surgery Department, Maternity and Children Hospital, 21955 Mecca, Kingdom of Saudi Arabia
Tel: +966 598 759 063; fax: +966 125 530 400;
e-mail: maher_zaiem@hotmail.com

Received 29 March 2014 **accepted** 26 May 2014

Introduction

Segmental dilatation of the intestine is a rare condition characterized by localized dilatation of the intestine and an abrupt transition between the normal and the dilated segments. It was first reported by Swenson and Rathauser in 1959 [1].

Segmental dilatation of the intestine can affect any part of the gastrointestinal tract from the duodenum to the rectum. Congenital segmental dilatation of the colon (CSDC) is a rare entity; it has been described mostly in children [2]. It is even more rare in neonates [3].

Mahadevaiah *et al.* [4] reviewed the literature in 2011 and found only nine cases of CSDC reported in neonates.

The characteristic features of CSDC include abrupt transition to the normal proximal and distal colon [5], hypomotility of the dilated segment, the absence of tenia coli, and abundant serosal vascularization fed by a large, marginal colonic artery [6].

We report a case of CSDC, presented in the neonatal period with a clinical picture of intestinal obstruction similar to that of Hirschsprung's disease, diagnosed preoperatively by contrast enema, and treated surgically by resection of the dilated segment and the Soave pull-through procedure. We discuss the clinical features, the radiological picture, the histopathological findings, and the surgical modalities for this pathology.

Case report

A full-term male baby, delivered by spontaneous vaginal delivery, with a birth weight of 3 kg presented on the first day of life with complaints of decreased oral intake, lethargy followed by abdominal distension. There was no history of vomiting and the baby passed meconium spontaneously.

On examination, he was afebrile but lethargic and mildly dehydrated. The abdomen was moderately distended and mildly tense, but there was no tenderness on palpation. An abdomen radiograph revealed a hugely dilated colonic shadow. A rectal tube was passed, and a moderate amount of well-formed stool was drained along with a large gush of air, and immediately the abdominal distension decreased significantly.

The patient was admitted in neonatal ICU for further management. During the hospital course, the patient was managed with frequent rectal washes along with supportive medical management. Meanwhile, the patient also kept on passing spontaneous stool. Contrast enema was performed, and it revealed a hugely dilated recto-sigmoid colon with abrupt normal-caliber colon proximal to it and without any transition zone (Fig. 1). The proximal end of the dilated segment was at the lower part of the descending colon, and the distal end of the dilated segment was at the level of the third sacral vertebra. This radiological finding was suggestive of segmental dilatation of the recto-sigmoid colon. Hence, surgery was planned, and we started by laparoscopy, which helped visualize the segmental dilation of the colon along with its prominent vasculature (Fig. 2). Laparotomy was performed, and the macroscopic appearance of the dilated colonic segment showed abundant serosal vascularization with a large marginal colonic artery, and the tenia coli was absent (Fig. 3).

Resection of the dilated segment was performed, and then Soave pull-through of the normal-caliber colon was performed and anastomosed to the anus (Fig. 4). The resected segment measured 20 cm in length and 15 cm in width.

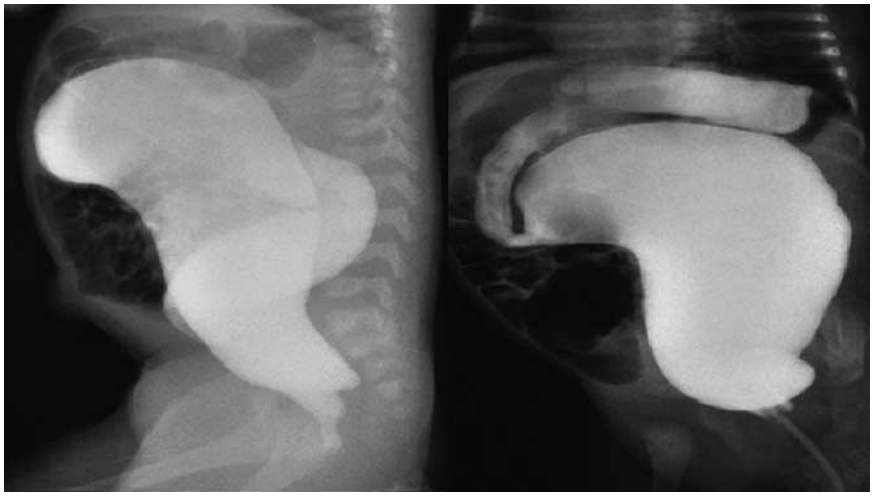
The postoperative course was smooth; feeding was resumed on the fourth postoperative day, and the baby was discharged on the eighth postoperative day in stable condition.

Histological examination demonstrated the presence of ganglion cells on both the submucosal layer and the muscularis propria of the dilated resected segment. Nerve fibers were hyperplasic and hypertrophic. In the muscularis propria layer, there was significant disarrangement, disorganization, and focal multiple atrophy of muscle fibers.

The presence of significant dilatation of blood vessels was also noted in the subserosal layer. The ganglion cells were present in the distal rectum.

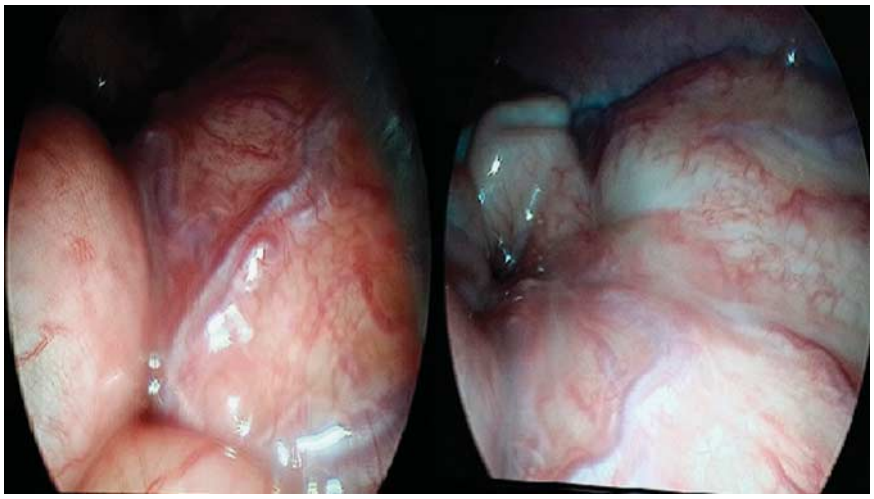
1687-4137 © 2015 Annals of Pediatric Surgery DOI: 10.1097/01.XPS.0000452059.63870.bc

Fig. 1



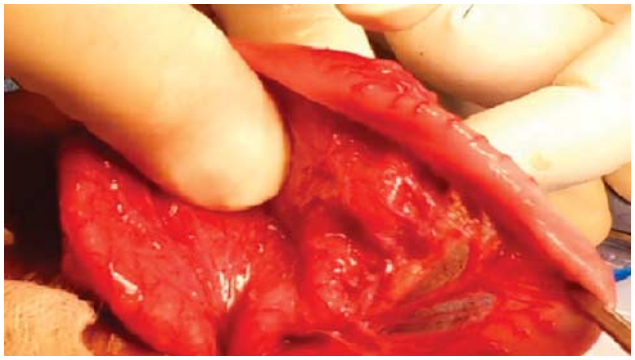
Contrast enema reveals a hugely dilated recto-sigmoid colon with abrupt normal-caliber colon proximal to it, without a transition zone.

Fig. 2



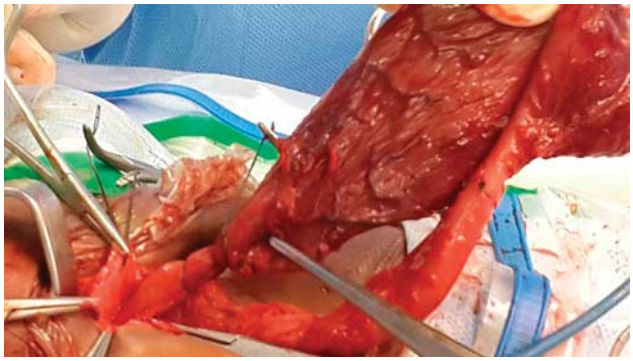
The laparoscopy view demonstrates the segmental dilation of the colon with prominent vasculature.

Fig. 3



The perioperative view shows a large marginal colonic artery, the prominent vasculature, and the normal colonic caliber proximal to the segmental dilatation.

Fig. 4



Endorectal Soave pull-through.

Discussion

Swenson and Rathauser in 1959 [1] were the first to describe three cases of segmental dilatation of the colon.

Since then, there have been several case reports of CSDC in the literature mostly described in children beyond the neonatal period [2,7].

Mahadevaiah *et al*. [4] reviewed the literature in 2011, and found nine cases of CSDC reported in neonates. Most of these neonatal cases were diagnosed peroperatively [8], whereas in our case, the diagnosis was made preoperatively due to the classical picture of the contrast enema, demonstrating an abrupt transition between the dilated segment and the normal colon proximally and distally; this radiological picture differentiates CSDC from Hirschsprung's disease.

It has been presumed that segmental dilatation of the colon is a congenital malformation [9,10]; the early presentation in the first day of life in our case supports this assumption.

The etiology of CSDC is still unknown. Several theories have been proposed to explain its pathogenesis. Mathé *et al*. [11] proposed a primitive neuromuscular dysfunction of the bowel, but this presumed theory does not explain the occurrence of the disease in a selective segment of the colon. In our case, two interesting findings were present in the histopathological examination: the first one is the presence of abundant, dilated, and tortuous blood vessels noted in the mesentry and the subserosal and the seromuscular layers of the dilated segment, which may play a role in the pathogenesis of the disease; this feature was also noted by Al-Salem [12]. The second interesting histopathological finding in our case was the presence of significant disarrangement, disorganization of the muscle fibers in the muscularis propria layer, associated with focal multiple atrophy. The absence of muscle hypertrophy in our case complies with the hypothesis raised by Helikson *et al*. [9], who supposed that the muscle hypertrophy observed in the cases discovered in older infants is acquired.

The surgical treatment of CSDC consists of the resection of the dilated segment and re-establishment of the intestinal continuity by end-to-end anastomosis [8]. Al-Salem [12] performed a staged Duhamel's pull-through after thorough bowel preparation. In our case, the distal end of the dilated segment was just at the level of the pelvic peritoneal reflexion. Hence, we performed the Soave pull-through technique, which is proved to be easy in the neonatal period, requiring minimal pelvic dissection with minimal danger of injuring the surrounding pelvic structure and achieving a safe extra-abdominal recto-anal anastomosis. An alternative technique is the resection of the dilated segment with an end-to-end anastomosis. This, however, requires a pelvic dissection with the intra-abdominal anastomosis.

Conclusion

CSDC, although a rare condition, should be considered in the differential diagnosis of neonatal low-intestinal obstruction. Barium enema is diagnostic, and differentiates CSDC from Hirschsprung's disease and other causes of intestinal obstruction in the neonatal period.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

References

1 Swenson O, Rathauser F. Segmental dilatation of the colon: a new entity. *Am J Surg* 1959; **97**:734–738.
2 Al-Salem AH, Grant C. Segmental dilatation of the colon. Report of a case and review of the literature. *Dis Colon Rectum* 1990; **33**:515–518.
3 Yadav K, Singh G, Budhiraja S. Congenital segmental dilatation of intestine. *Indian J Pediatr* 1996; **63**:561–563.
4 Mahadevaiah SA, Panjwani P, Kini U, Mohanty S, Das K. Segmental dilatation of sigmoid colon in a neonate: atypical presentation and histology. *J Pediatr Surg* 2011; **46**:e1–e4.
5 Brawnier J, Schafer D. Segmental dilatation of the colon. *J Pediatr Surg* 1973; **8**:957–958.
6 Takehara H, Komi N, Hino M. Congenital segmental dilatation of the colon: report of a case and review of the literature. *Pediatr Surg Int* 1988; **4**:66–68.
7 De Lorimer AA, Benzian SR, Gooding CA. Segmental dilation of the colon. *Am J Roentgenol* 1971; **112**:100–104.
8 Sarin YK, Singh VP. Congenital segmental dilatation of colon. *Indian Pediatr* 1995; **32**:116–118.
9 Helikson MA, Schapiro MB, Garfinkel DJ, Shermeta DW. Congenital segmental dilatation of the colon. *J Pediatr Surg* 1982; **17**:201–202.
10 Chiba T, Kokubo T. Congenital segmental dilatation of the colon. *Nihon Geka Hokan* 1976; **45**:45–47.
11 Mathé JC, Khairallah S, Phat Vuoung NP, Boccon-Gibod L, Rey A, Costil J. Dilatation segmentaire du grêle à révélation néonatale. Etude par imprégnation argentique des plexus myentériques [Segmental dilatation of the ileum in a neonate. Study of the myenteric plexus with a silver staining preparation]. *Nouv Presse Med* 1982; **11**:265–266.
12 Al-Salem AH. Congenital segmental dilatation of the rectosigmoid colon: a forgotten cause of constipation. *J Pediatr Surg Spec* 2008; **2**:20–22.