Acta Paediatrica Hungarica, 25 (4), pp. 377—383 (1984)

Congenital Short Colon Associated with Imperforate Anus (Zachary-Morgan syndrome)

J Dénes, K Ziszi, M Bognár, E Schläffer, J Tóth

Apáthy Children's Hospital, Budapest, Hungary

Two cases of short colon with imperforate anus are presented. Terminal colostomy is the therapy of choice in the neonatal period. As the definitive final procedure, trimming into a tubular structure and abdominoperineal pull-through operation is suggested. A survey is given of similar cases reported in the literature.

Among the anomalies of the colon, besides Hirschsprung's disease atresia is the best known condition, although it is ten times less frequent than atresia of the small intestine, which occurs about once in every 15 000 to 30 000 births [7]. Neonatal small left colon syndrome, reported first by Davis et al in 1974, is similarly rare [3]. We found only 3 cases of total agenesis of the large intestine in the literature [1, 5, 6], and the so-called "short colon with imperforate anus" or Zachary—Morgan syndrome, has only been published in 8 papers [2, 4, 8, 9, 10, 12, 13, 14]. This malformation was first described by Trusler et al in 1959 [12] who reported 7 such cases. Later 7 other authors published similar cases, generally 1 or 2, except for two publications from India [8, 9], in which 10 and 6 cases respectively were presented. It is interesting that the two hospitals where these patients were observed, are only 70 km distant, both in the province of Punjab. In these two papers boys were more frequently affected than girls: in one paper the ratio was 8 to 2, in the other, 4 to 2. In the other publications the numbers of boys and girls were nearly the same. Trusler called this malformation short colon; since Zachary' and Morgan's report [11] some authors call it Zachary—Morgan syndrome.

The malformation is characterized by the absence of the left half of the colon together with a saccular dilatation and imperforate anus. Colovesical, colovaginal or coloperineal fistula is often but not always present. The terminal ileum is usually entering at the upper quadrant of the dilated colon, which is occupying the greater part of the abdomen. There is no taenia to be found on the thick, hypertrophied and distended colon and histological examination shows normal ganglion cells in its wall. The short colon is supplied by branches of the superior mesenteric artery, the inferior mesenteric artery is usually missing. The aetiology of the anomaly is unknown; Dickinson [4] suggested that it results from an occlusion or

nondevelopment of the inferior mesenteric artery very early in fetal life. As associated malformations, first of all Meckel diverticulum, malrotation, hydronephrosis, uterus bicornis and hypospadias were described.

Reports of Cases

Case 1. K. M., a full-term baby boy, weighing 3050 g, was admitted with imperforate anus and abdominal distension. On the plain roentgenogram high anorectal atresia was to be seen with large air-fluid level (Fig. 1). A temporary loop

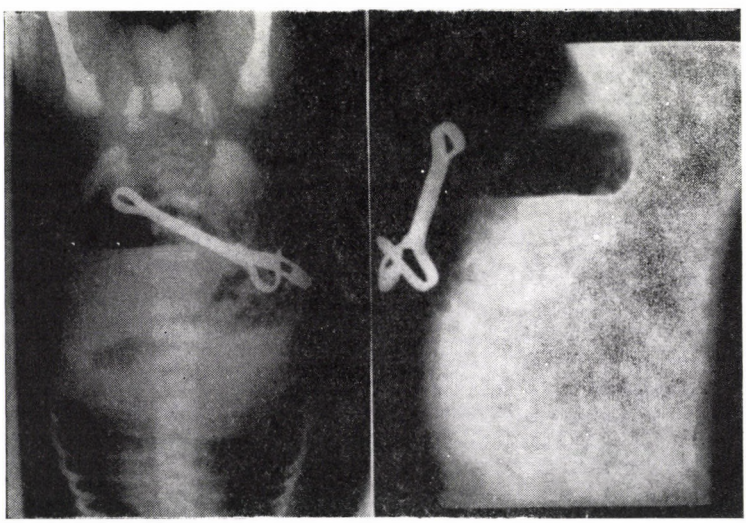

Fig. 1. Plain roentgenogram showing large air-fluid level (Case 1)

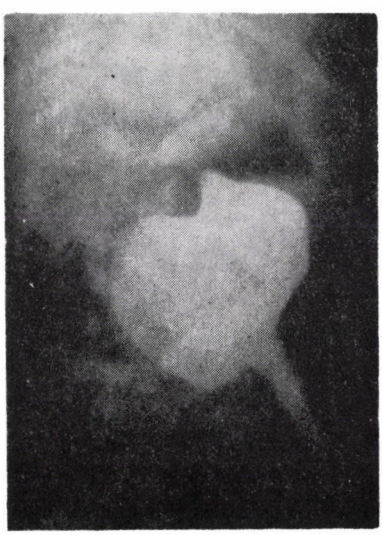

Fig. 2. Contrast study of the saccularly dilated colon (Case 1)

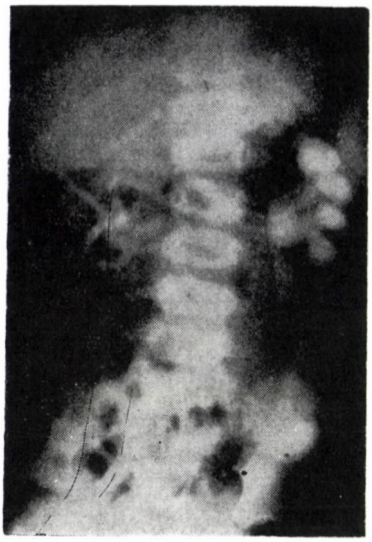

FIG. 3. Urogram showing hydronephrosis on left side (Case 1)

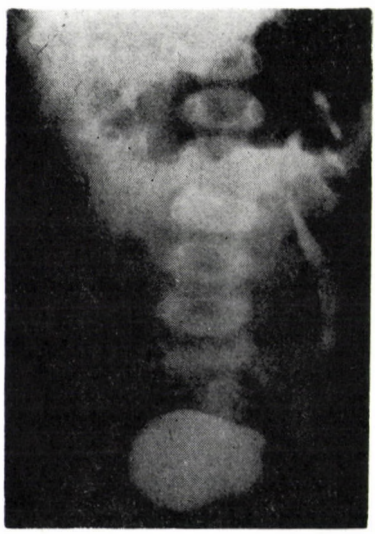

FIG. 4. Urogram after surgery (Case 1)

colostomy was planned, but on opening the abdomen instead of the normal large intestine, a great saccular dilated segment of colon was found with upper entering of the terminal ileum without any fistula into the bladder. Hence a terminal window colostomy was performed, which did not function well later and needed revision: widening the stoma because of stenosis. After the widening repeated prolapses occurred, so we had to fix the wall of the sac to the abdominal wall to prevent further prolapse. The saccular dilatation of the colon could be identified by contrast study (Fig. 2). The intravenous urogram showed a normal situation on the right side, but hydronephrosis of the left kidney (Fig. 3). On exposure of the left ureter we

found a 2 cm long fibrous stricture next to the distended colon. After resection and end-to-end anastomosis of this part of the ureter, the hydronephrosis regressed gradually (Fig. 4). The colon failed to decrease in size and showed poor motor function retaining fecal material, so we had to evacuate it manually from time to time, with the aid of repeated enemas, to keep the colon decompressed. 10 months later, when the child was one year old, as a final definitive procedure, we resected most of the ganglionic but not functioning colon and made an abdomino-perineal pull through operation with the terminal ileum and caecum (Fig. 5). The child now is two years old, developing quite well and having 3—4 stools a day.

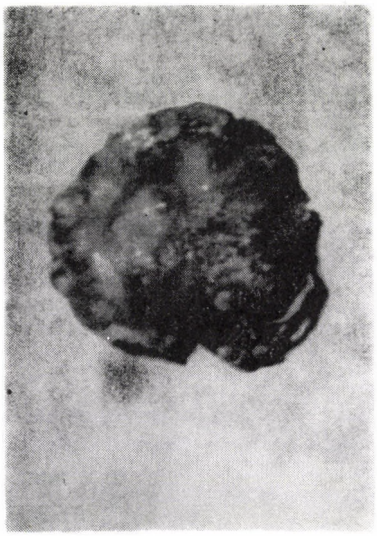

FIG. 5. Resected colon sac (Case 1)

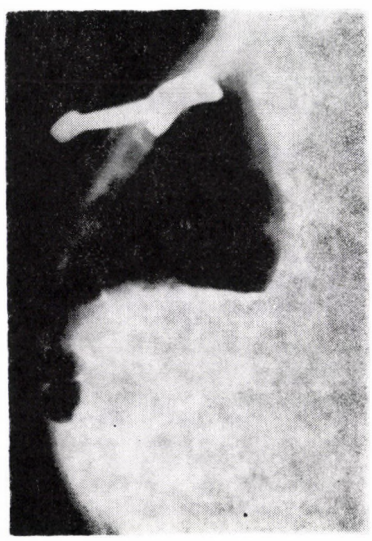

FIG. 6. Plain roentgenogram showing large air-fluid level (Case 2)

FIG. 7. Contrast study of the saccularly dilated colon (Case 2)

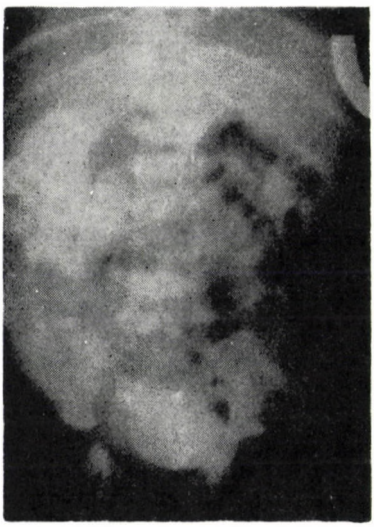

FIG. 8. Urogram showing hydronephrosis on left side (Case 2)

Case 2. M. K. a full-term baby girl weighing 3500 g was admitted with imperforate anus, distended abdomen and a later identified Down syndrome. The plain roentgenogram showed a large air-fluid level (Fig. 6). On opening the abdomen we found a large distended colon segment similar as in the first case. Terminal colostomy was performed which later needed repeated dilatations. Contrast study showed a saccular dilatation of the short colon (Fig. 7). On the intravenous urogram enlargement of the left kidney was to be seen with distended calyces and decreased excretion (Fig. 8). One month later the excretion seemed to have improved on the urogram. The infant is now one half year old, developing well and as

TABLE I

Main clinical features of the reported cases

Year	Author	Number of cases	Sex	Fistula	Treatment	Result
1959	Trusler et al	7	3 boys 4 girls	3 colovesical 2 colovaginal	3 colostomies 1 ileostomy	3 survived 4 died
1962	Zachary Morgan	2	2 girls	1 colovesical 1 colovaginal	1 colostomy 1 ileostomy	2 survived
1967	Dickinson	1	boy	Ø	ileostomy	died
1967	Singh A et al	10	8 boys 2 girls	5 colovesical	9 colostomies 3 clos. fist. 1 pull through	7 survived 3 died
1967	Shafie	1	girl	coloperineal	anoplasty colostomy	died
1972	Singh S Pathac	6	4 boys 2 girls	3 colovesical 1 colovaginal	5 colostomies 1 ileostomy	3 survived 3 died
1976	Chiba et al	2	?	?	1 colostomy 1 pull through colostomy	2 survived
1982	Vaezzadek et al	1	girl	colovaginal	clos. fist. pull through	survived
1983	Dénes et al	2	1 boy 1 girl	Ø	2 colostomies 1 pull through	2 survived

a final definitive procedure we are planning trimming of the colon into a narrower tubular structure. This is hoped to result in improved motor function and emptying. We plan to bring it down with an abdomino-perineal pull-through.

DISCUSSION

In both our cases we found a typical saccular dilatation of the short colon with imperforate anus without fistula. As associated malformations, hydronephrosis was present in both children and Down syndrome in the second one. Because of the high anorectal obstruction, terminal colostomy had to be performed in both newborn babies. In the 30 cases reported until now, 21 colostomies and 4 ileostomies were done and 2 dilatations of the vaginal and perineal fistulas. Disconnection and closure of the colovesical and colovaginal fistulas were carried out in 4 cases. In 3 infants abdomino-perineal pull through operations were the final procedure, once with resection of the whole sac and three times with trimming. In our first case we made a pull through after subtotal resection of the sac, but in the second one we are planning trimming to a tubular structure if the sac fails to decrease in size. Of the reported 30 cases 18 survived, 12 died. In most fatal cases associated malformations have been responsible

for the mortality. Main clinical features of the reported cases are given in Table I.

There is every reason to hope that with better diagnosis of this rare anomaly and with improvement of the operative procedures, the prognosis and the results of therapy will be more promising.

References

1. Blunt A, Rich GF: Congenital absence of the colon and rectum. Am J Dis Child 114:405, 1967
2. Chiba T, Kasai M, Asakura Y: Two cases of coloplasty for congenital short colon. Arch Jpn Chir 45:40, 1976
3. Davis WS, Allen RP, Favara BE: Neonatal small left colon syndrome. Am J Roentgenol Radium Ther 120: 322, 1974
4. Dickinson SJ: Agenesis of the descending colon with imperforate anus. Am J Surg 113:279, 1967
5. Gerner L, Tison P: Agenesis of large intestine in a newborn infant with anal imperforation. Am Anat Path 16:370, 1939
6. Klinefelter EW: Congenital absence of the colon. Am J Dis Child 50:454, 1935
7. Powel RW, Raffensperger JC: Congenital colonic atresia. J Pediatr Surg 17:166, 1982
8. Singh A, Singh R, Singh A: Short colon malformation with imperforate anus. Acta Paediatr Scand 66:589, 1967
9. Singh S, Pathac IC: Short colon associated with imperforate anus. Surgery 71:781, 1972
10. Shafie ME: Congenital short intestine and cystic dilatation of the colon, associated with imperforate anus. J Pediatr Surg 6:76, 1971
11. Tosovsky V: Nahle prihody brisni u deti, ed. Avicenum, Praha 1981, p. 109
12. Trusler GA, Mastel AL, Stephens CA: Colon malformation with imperforate anus. Surgery 45:328, 1959
13. Vaezzadek K, Gerami S, Kalani P, Sieber WK: Congenital short colon with imperforate anus: a definitive surgical cure. J Pediatr Surg 17:198, 1982
14. Zachary RE, Morgan A: Prvostrevo. Cs Pediat 17:208, 1962

Received December 15, 1983

J Dénes MD,
Bethesda 3
1146 Budapest, Hungary