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Pediatric Radiology

Radiological findings in total aganglionosis coli

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Abstract. The radiological findings in 13 patients with total aganglionosis coli were reviewed. There was a male to female ratio of 7:6. Fifty-four percent of patients presented in the first week of life, but a significant number (31%) did not present until after 1 month of age. All patients had plain film evidence of bowel obstruction when referred for a barium enema. There were no pathognomonic barium enema findings, and barium enema results covered the entire spectrum of findings which can be encountered in the neonate and young infant with bowel obstruction. Seventy-seven percent had normal calibre colon, 23% had micro colon, 23% had a shortened colon, 46% had colonic wall irregularity, 33% had significant ileal reflux. Delayed evacuation of barium from colon occurred in the two patients who had delayed films. Total colonic aganglionosis should be considered in any infant or young child with plain film evidence of bowel obstruction, whatever the barium enema findings. Hirschsprung's disease and the level of transition can only be definitively diagnosed by biopsy.

The purpose of this study was to determine the radiological findings in total aganglionosis of the colon.

The literature on the radiological findings in total aganglionosis coli (TAC) is contradictory. Sane et al. [13] reviewed 76 patients from the literature and found that 38% had microcolon, 21% had normal barium enemas, 21% had prolonged retention of contrast, 19% had a short colon of normal calibre, 15% had apparent colonic transition zones, 8% had meconium plugs, 5% had barium reflux into small intestine, 4% had abnormal contractions and 2% had megacolon. In his series, the term microcolon included "any colon whose lumen is of manifestly small calibre", and was not confined to the pencil thin colon associated with meconium ileus. Berdon et al. [1, 2] reported six patients with TAC who had colons of normal calibre but appeared shortened with loss of redundancy in their sigmoid loop, splenic and hepatic flexures. None had microcolons.

Cremin et al. [6] also reported the radiological features of six patients with TAC. He emphasized the findings of rapid reflux of contrast into the terminal ileum, discrepancy of size between normal colon and enlarged small bowel loops resulting in a relatively narrow colon, retention of barium in the entire colon and loss of the normal redundancy of flexures resulting in a shortened appearance of the colon. Chandler et al. [5] described three cases of TAC, of which two demonstrated retrograde filling of the entire small bowel.

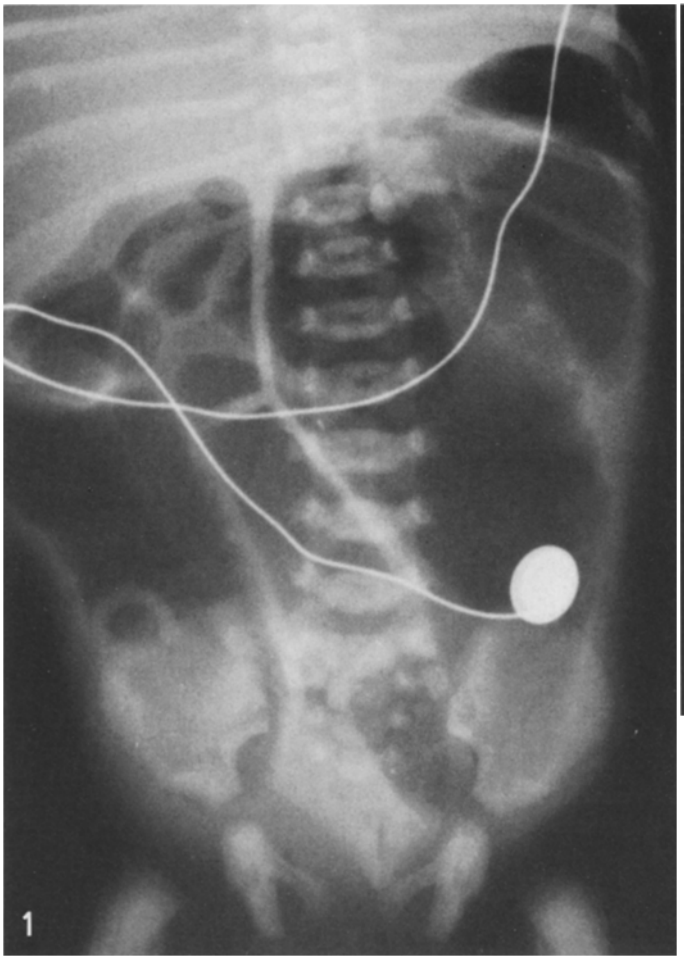
Materials and methods

Between 1954 and 1981, 27 cases of aganglionosis of the entire colon with variable involvement of small bowel were treated at the Royal Children's Hospital [4].

The diagnosis of Hirschsprung's disease was made by serial paraffin sections on suction rectal biopsies [3], and since 1975 also by histochemical estimations of acetylcholinesterase [11]. The extent of disease was determined by frozen section at the time of laparotomy, and confirmed subsequently by examination of the resected aganglionic bowel.

It is current practice in our Institution for the diagnosis of Hirschsprung's disease to be made by histochemical demonstration of acetylcholinesterase on suction rectal biopsies and confirmed with paraffin sections. Colostomies are sited after frozen section confirms the presence of ganglion cells. Manometry is not used.

Thirteen enemas were available for review. The enemas were reviewed and evaluated for criteria mentioned in the literature [1, 2, 4–10, 12–14]. Patients who had TAC and contrast enemas but who did not have the enemas available for review were not in-



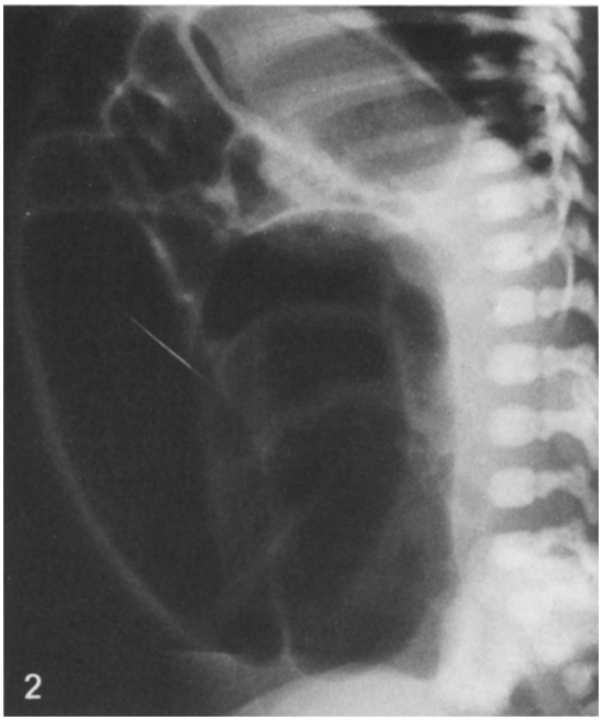


Fig. 1. A plain abdominal film of a 2-day-old infant with multiple distended bowel loops; typical of the plain film findings of infants with total aganglionosis coli at the time of presentation for barium enema

Fig. 2. A vertical beam lateral abdominal film of a 2-day-old infant with total aganglionosis coli. Note the relatively undistended rectum with distended bowel loops elsewhere with aparent transition of rectosigmoid function

Table 1. Age of patients when barium enema performed

Age of patients	Number	Percentage
Less than 3 days	4 patients	31
4 to 7 days	3 patients	23
One week to one month	2 patients	15
Greater than one month	4 patients	31

cluded in the study. The diagnosis of microcolon was restricted to patients with pencil-thin colon of the type normally associated with meconium ileus. Evaluation of colonic length (colonic redundancy) and mucosal irregularity was necessarily subjective.

Results

Sex and age

Plain films and contrast enemas were available for review in 13 patients with aganglionosis of the entire colon. Six were female and seven were male. The age distribution of patients at the time barium examination was performed is summarized in Table 1.

Plain film findings

At the time of barium examination, all patients had plain film findings of bowel obstruction (Fig. 1) with multiple gas distended loops of bowel with multiple fluid levels. Meconium was detected on the plain films of two patients, simulating the findings in meconium ileus. Plain film findings were suggestive of short segment Hirschsprung's disease in two patients in whom the lateral views of the rectum showed a distended sigmoid colon apparently continuous with a narrow rectum (Fig. 2).

Barium enema findings

Of the 11 patients who had normal calibre colons, 8 had colons of normal length and 3 had shortened colons. Five of these 11 patients showed irregularity of the colonic wall consistent with "spasm" (Fig. 3).

Fig. 3. Abdominal film obtained during barium enema shows normal colonic calibre, meconium filling defects, irregular colonic wall consistent with spasm and loss of the normal sigmoid redundancy.

Fig. 4. Barium enema showing microcolon in a 2 day old infant with total aganglionosis coli.

Of these 5, 2 had normal length colons and 3 had shortened colons.

Two patients with microcolons both had normal colonic length (Fig. 4). One had an irregular bowel contour suggestive of "muscle spasm". Both were examined in the first 3 days of life.

Of the 2 patients suspected on plain films of having short segment Hirschsprung's disease, 1 had this suspicion confirmed by barium enema. No patient had the findings of colitis. Meconium plugs were present in 7 patients and none of these had microcolon (Fig. 5). 24-h delayed films were obtained in 2 patients and barium was present throughout the colon in both (Fig. 6). Significant reflux of contrast into the terminal ileum occurred in four patients (right side of colon was not opacified in 2 patients) and in two of these the opacified ileum was markedly distended.

Discussion

As with previous studies, the incidence of TAC in our study approaches an even distribution between males and females, and differs from the male predominance (4 to 1) seen in the usual short segment Hirschsprung's disease [12, 13].

Many authors have emphasized the lack of correlation between severity of symptoms, the age of presentation and the extent of colonic involvement in Hirschsprung's disease [4, 12]. In our 13 patients with TAC the age of presentation was quite variable. While 54% presented in the first week of life, 31% did not present until after four weeks of age. In their review of all cases of total anganglionosis at our institution between 1954 and 1982, Cass and Myers [4] found that mortality has greatly decreased over the period. This appears related to earlier diagnosis of Hirschsprung's disease and its full extent, rather than changes or improvements in surgical technique.

All patients in our series had plain film evidence of bowel obstruction at the time they were referred for barium enema. Thus TAC enters into the differential diagnosis of bowel obstruction in the first three months of life. Misleading plain film findings of meconium ileus (2 patients) and of short segment Hirschsprung's disease (2 patients) served to empha-

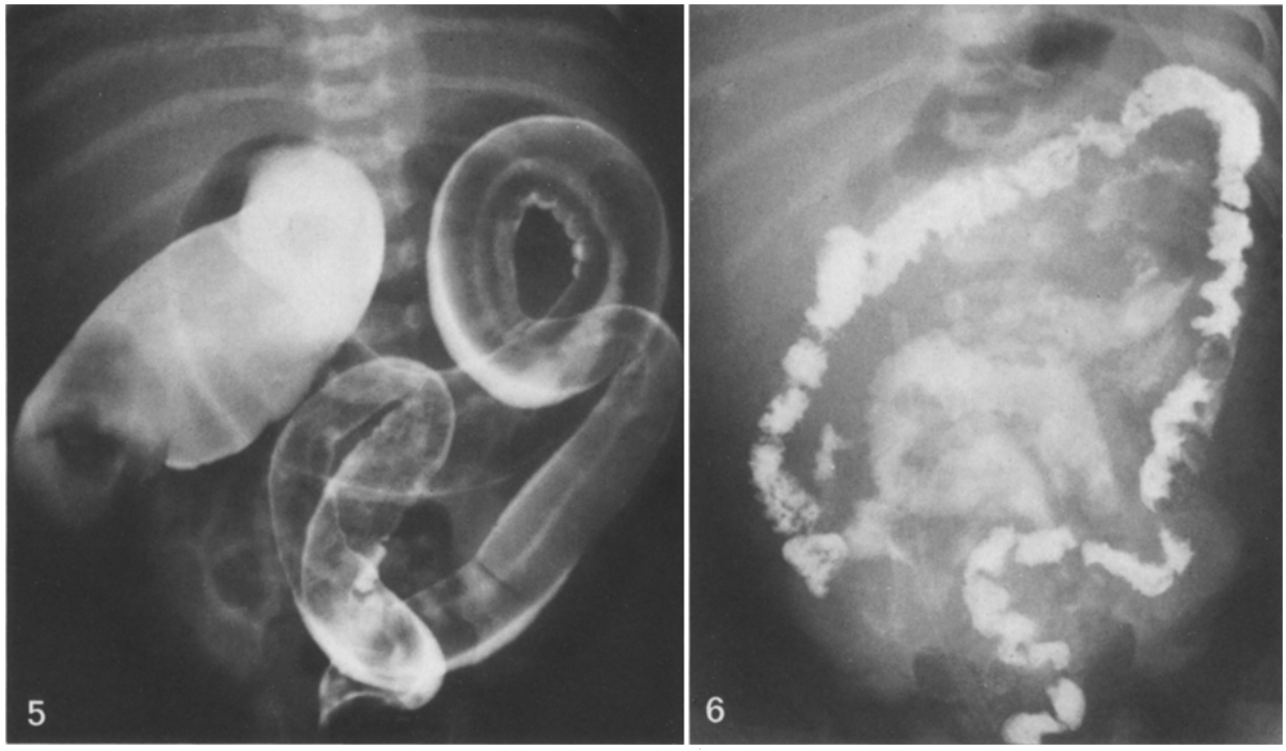


Fig. 5. Barium enema in an infant showing large meconium plugs in a colon with normal redundancy of flexures and sigmoid, and normal calibre colon.

Fig. 6. A 24 hour delay film after a barium enema showing retained barium in distended small bowel loops and collapsed colon.

size the difficulties in distinguishing between large and small bowel in plain films performed on neonates and young children [14].

Our barium enema results confirmed that there are no pathognomonic signs of TAC. The findings at barium enema covered the spectrum of results which can be encountered in the neonate and young infant with bowel obstruction. Of all the barium enema findings studied, colonic calibre was the easiest to evaluate. In 85% of cases the colon was of normal calibre, and the remainder had a microcolon. The lesser incidence of microcolon in this series may reflect the stricter criteria applied for its diagnosis. TAC should be considered in the differential diagnosis of neonates with bowel obstruction and (a) a barium enema showing a colon of normal calibre along with jejunal atresia, meconium plug syndrome, and short segment Hirschsprung's disease; (b) in the differential diagnosis of microcolon along with ileal atresia, low jejuneal atresia and meconium ileus.

Colonic length was judged to be shortened, with loss of normal redundancy of sigmoid colon, splenic and hepatic flexures in 23% of patients. This is similar to the findings of Sane [13]. The difficulty in evaluating this subjective sign and its relative infrequency make this an unreliable sign in the diagnosis of TAC unless other findings are present.

Meconium plugs were present in 54% of patients, but this is also a nonspecific finding since these may be present in normal children, the meconium plug syndrome, meconium ileus and short segment Hirschsprung's disease [10]. The single patient with a pseudotransition zone in the rectosigmoid emphasises the need for biopsy confirmation of the transition zone [7, 12], complete enema and delayed films [9, 14].

Other ancillary signs which can be of use in raising the suspicion of TAC are colonic wall irregularity (46% of our patients), and gross ileal reflux (36% of our patients). Although some authors have found this latter sign rarely, Cremin [6] and Chandler [5] have found this consistently in their studies of 6 and 3 patients respectively.

Retention of barium at 24 h is regarded by some [2, 8] as the only pathognomonic sign of TAC so there should be routine delayed films after barium enema regardless of the provisional diagnosis at the conclusion of the enema if these children are not proceeding directly to laparotomy. In this way, the diagnosis of TAC will not be overlooked. Only two

of our patients had delayed films, but both had retained contrast throughout the entire colon. However, the reliability of this finding is questioned by French [7]. Thus a complete enema to determine the position of the caecum (to exclude malrotation), to detect barium reflux into the distended small bowel, and a 24-h film should be obtained in all neonates and young infants with bowel obstruction.

Conclusion

Hirschsprung's disease should only be diagnosed by histological or histochemical examination. Radiological estimates of the transition zone serve only as a guide to the initial choice of site for colostomy/ileostomy, and presence of ganglion cells must be confirmed by frozen section.

There are no pathognomonic barium enema findings in TAC. TAC should be considered in a differential diagnosis of all neonates and young infants who present with radiological evidence of bowel obstruction whatever the barium enema findings. Although the majority of patients have a normal calibre colon some have a microcolon. The index of suspicion for this disease should be raised if the colonic wall is irregular, gross ileal reflux occurs into distended small bowel, the colon has lost normal redundancy of the flexures, meconium plugs are present or there is delayed evacuation of barium from the entire colon at 24 h. Neonatal barium enemas should be complete and delayed films always obtained.

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