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Total colonic aganglionosis: 30 years' experience

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Abstract. Thirty-two patients with total colonic aganglionosis (TCA) were seen in 30 years, representing 10% of the total number with aganglionosis seen during that period (1954–1983). The mortality rate was 45%, but has shown a progressive decrease from 82% in the first decade to 9% in the last. The improvement is specifically related to earlier diagnosis, control of sepsis, and the use of parenteral nutrition in dealing with ileostomy losses, but appears to have been independent of the definitive surgical procedure. Follow-up study indicates some delay in achieving continence without any clear-cut relationship to the operative procedure. The 32 patients have been reviewed with particular emphasis on sex ratio, age at diagnosis, clinical features, and results of treatment.

Key words: Hirschsprung's disease – Total colonic aganglionosis

Introduction

Total colonic aganglionosis (TCA) occurs in 2–13% of patients with Hirschsprung's disease [2, 8, 9, 14, 19, 22, 23, 36, 38]. This subgroup of Hirschsprung's disease presents particular problems of diagnosis and treatment, with early series reporting mortalities of 65% or more [2, 13, 14, 40]. In more recent series, the mortality has decreased to 0–25% [1, 6, 19, 25]. This decrease has been attributed to earlier diagnosis and better support [6] or to improvements in operative technique [24, 25]. There is still debate as to the optimal surgical technique, with several new operative variations having been reported recently [3, 20, 25, 29, 33].

In this paper, 32 patients treated over a 30-year period are reviewed in order to assess changing patterns of treatment and to compare the results of different operative procedures.

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Clinical series

The first documented case of TCA in this series was in 1954. In the initial patients, diagnosis was late and mortality high, the first survivor occurring in 1957. In the 30 years from 1954–1983, there were 32 patients with TCA among a total of 305 patients with Hirschsprung's disease (10%). Twenty-four were males (75%). One infant was pre-term (35 weeks gestation); the gestational period was between 37 and 40 weeks in 6; 40 weeks in 22; and more than 40 weeks in 3. Birth weights ranged from 2.4–4.8 kg, with a median of 3.4 kg. Two patients had associated anomalies: one had Morquio's disease and the other transposition of the great vessels with asplenia.

Five patients (15%) had a family history of Hirschsprung's disease; 2 were father and son, 3 had a relative with a transition zone in the rectosigmoid. One of these patients had a grandmother and maternal niece diagnosed in adult life and 2 had an affected sibling (one of these siblings also developed a fatal neuroblastoma at 3½ years of age).

Twenty-five patients presented in the neonatal period; 23 of these were under 1 week of age. Seven patients presented beyond the neonatal period, the oldest at age 4 months. Four of these 7 patients had neonatal symptoms, while 3 had none. Presenting features were vomiting and distension in 29 patients (90%) and failure to pass meconium in 16 (75%) of the 21 patients in whom this information was recorded. In infants presenting after 1 month of age, severe constipation was the predominant symptom. There was a delay in diagnosis in 16 patients, which for 8 was a contributing factor to their death. Even in recent years there has been diagnostic delay, and the difficulties in diagnosis are illustrated by the following case histories.

Case 28. This infant commenced bile-stained vomiting at 6 h of age. An abdominal X-ray at 20 h of age showed multiple

fluid levels felt to be consistent with necrotising enterocolitis. A repeat abdominal X-ray at 36 hours showed free gas. At laparotomy, a diagnosis was made of necrotising enterocolitis with colonic perforation and a transverse colostomy was performed just proximal to the site of perforation. Histological analysis of the specimen from the colostomy showed aganglionosis. At repeat laparatomy on day 12, ganglion cells were found 10 cm from the ileocaecal valve and an ileostomy was fashioned.

Case 32. This infant presented at 24 h with faeculent vomiting, distension, slight abdominal tenderness, and failure to pass meconium. An abdominal X-ray showed bowel obstruction with multiple fluid levels. Barium enema showed microcolon. Laparotomy revealed a volvulus of grossly distended small bowel which suddenly narrowed, after which there was collapsed small bowel containing firm, pale, inspissated bowel contents. At the base of the vovulus there were three perforations of the collapsed small bowel with localised peritonitis. Complicated meconium ileus was diagnosed an an ileostomy performed at the end of the dilated small bowel (45 cm from the ileocaecal valve). Histological examination revealed aganglonosis. Subsequent laparotomy demonstrated ganglion cells a further 5 cm proximally, and the ileostomy was resited.

Results

The overall mortality was 45%, but there has been a progressive improvement over the decades with the mortality dropping from 82% to 9% (Table 1). The principal cause of death was delay in diagnosis and treatment, with sepsis and nutritional problems being secondary (Table 2). In 8 of the 15 deaths there was a delay in diagnosis or inappropriate treatment. Additional complicating factors were a small bowel thrombosis in 1 patient who died and an associated anomaly in another. One patient (also initially misdiagnosed) died primarily of nutritional and fluid and electrolyte problems associated with ileostomy dysfunction. Of the 4 postoperative deaths, 2 patients had an early Swenson procedure which was followed by late sepsis in 1 and staphylococcal pneumonia in the other. Another postoperative death was due to enterocolitis 3 years after a Duhamel procedure. The most recent death, the only one in the past decade, occurred some months following a Duhamel operation, but necropsy indicated that death was unrelated to Hirschsprung's disease or the operative treatment.

Table 1. Mortality in 32 patients with TCA (divided into 3 decades)

Year	Patients	Deaths	Mortality
1954–1963	11	9	82%
1964–1973	10	5	50%
1974–1983	11	1	9%

Table 2. Cause of death in 15 patients with TCA

1. Before operation (4)
   Misdiagnosed, dyskinesia with sepsis
   Barium enema negative, died *Escherichia coli* septicaemia (also asplenic syndrome)
   Delay in diagnosis and transfer
   Aspirated and inhaled while waiting for laparotomy
2. Following laparotomy (7)
   Misdiagnosed, ileostomy for perforated ileum but fistulae after attempts to close ileostomy
   Missed diagnosis until moribund and second laparotomy
   Attempted conservative treatment but peritonitis from perforated caecal biopsy site
   Delayed diagnosis, small bowel thrombosis after ileostomy
   Ileostomy dysfunction and prolapse, tracheo-bronchitis at 5 months
   Delay and incorrect diagnosis, transverse colostomy followed by dehiscence
   Delayed diagnosis, moribund due to peritonitis associated with perforated stercoral ulcer in the transverse colon
3. Following definitive surgery (4)
   Early Swenson (day 2), leaked, failure to thrive, staphylococcal pneumonia, died at 3 months.
   Early Swenson (4 months), peritonitis at 8 months, post mortem revealed an intra-abdominal abscess.
   Enterocolitis 3 year after Duhamel operation.
   After Duhamel operation but unrelated.

While abdominal X-ray showed a distal bowel obstruction in 29 patients, the barium enema was less helpful. The radiology findings are summarised in Fig. 1 and barium enema findings in Table 3. Barium enema was performed in 16 patients and showed normal colon calibre and length in 9 patients; 3 patients had a "false cone" at the sigmoid colon. Two cases had reflux into the dilated terminal ileum, allowing a confident radiological diagnosis of TCA. In 2 cases there was active defaecation as soon as any large amount of barium was introduced. One patient who had a barium meal had passed all barium within 18 h. There were no pathognomonic features of TCA in the colon, but there were often suggestive features such as shortening of the colon and serrated irregular mucosa. The most important function of the barium enema, however, was to exclude other causes of neonatal bowel obstruction. The commonest finding in TCA was an essentially normal study.

The overall course and results of the 32 patients are shown in Fig. 2. The diagnosis of Hirschsprung's disease was made post mortem in 2 infants and a further 2 who were suspected of having Hirschsprung's disease died before operation could be undertaken. Of the 28 infants who

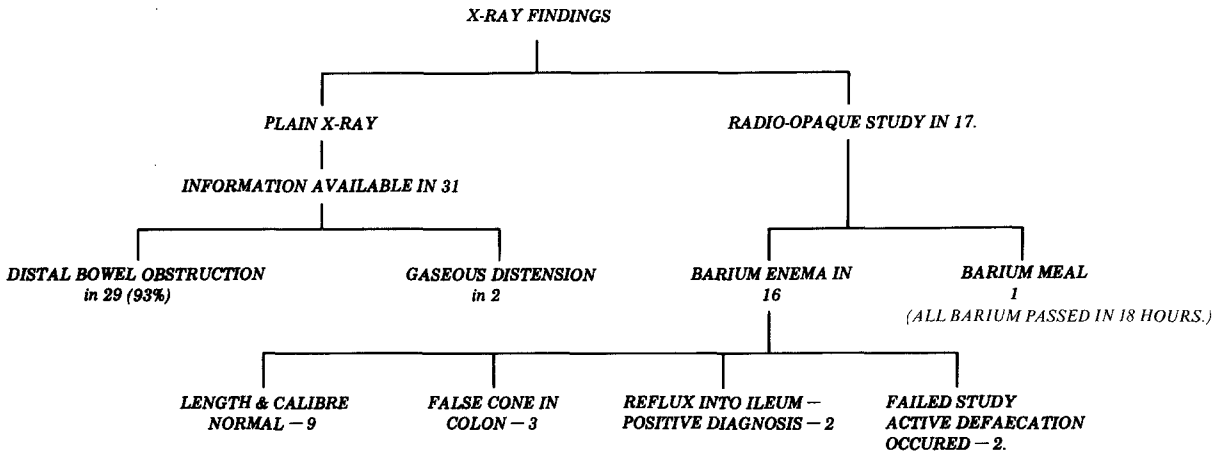

Fig. 1. Radiology findings in 31 of the 32 patients with total colonic aganglionosis

underwent surgery, only 5 had a pre-operative diagnosis of probable TCA. Sixteen patients had a pre-operative diagnosis of Hirschsprung's disease, the operative biopsies correctly determining the level of aganglonosis (although in 3 there was a macroscopic "cone" in the transverse colon). Four were diagnosed on the macroscopic findings at operation and had correct siting of the ileostomy. Two patients had initial "negative" laparotomies and 1 was misdiagnosed as complicated meconium ileus, requiring later resiting of the ileostomy. Five patients had initial incorrect transverse colostomies, 1 being misdiagnosed as necrotising enterocolitis because of a perforation and 4 because of a macroscopic cone in the transverse colon (making a total of 7 in whom there was a false cone). These diagnostic difficulties became less over the series with increasing use of pre-operative rectal suction biopsy, barium enema, and intra-operative frozen sections.

Table 3. TCA-summary of barium enema findings

Findings	Number	Percentage
Total patients	16	
Colonic calibre		
normal	12	75
megacolon	0	0
micro	4	25
Colonic length		
normal	12	75
short	4	25
Normal calibre and length	9	56
Mucosal irregularity		
normal	9	56
irregular	7	44
Ileal reflux	3/11	27
False transition zone	3	19
Meconium plugs	7	44
Colitis	0	0
Delayed evacuation	2/2	

The length of aganglionosis ranged from 3 cm to 120 cm from the ileocaecal valve (average 18 cm) (Fig. 3). Over the same 30-year period there were two cases of aganglionosis of the entire small bowel, and as there was no treatment possible in these cases, they were not included in the series.

The distal aganglionic bowel caused complications in 6 patients: 1 had was an intra-uterine vovulus with the perforations in the collapsed terminal ileum near the base of the twist. At resiting of the ileostomy this distal bowel was removed, as it contained copious amounts of pus. The second patient presented at 2 days of age with perforation of the transverse colon, the appearance mimicking necrotising enterocolitis. The third presented at 2½ months with a stercoral perforation of the transverse colon. The fourth and fifth patients died a few days after ileostomy from perforation of the colonic biopsy site (caecal and sigmoid). One of these patients had presented at 15 days of age with pus in the right groin, probably from perforation of the appendix. The sixth patient had the aganglionic bowel left in situ after a Swenson procedure. He developed an appendicular abscess, appendicular-vesical fistula, and recurrent urinary tract infections which persisted until the aganglionic bowel was removed.

Twenty-eight patients had ileostomies fashioned and there were 7 deaths early in the series. Five of these were due to sepsis in pre-operative moribund patients and 2 to late nutritional fluid and electrolyte problems. Ileostomy complica-

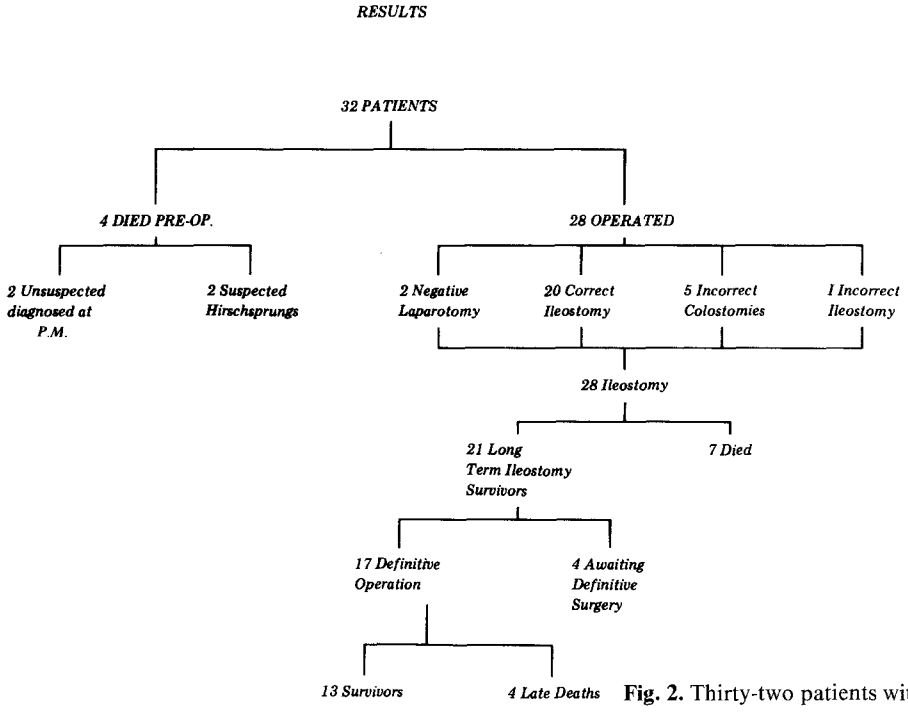

Fig. 2. Thirty-two patients with TCA: course and results of treatment

tions included 5 patients who had a prolapse and 6 with excessive losses requiring frequent hospital admissions and parenteral feeding. These excessive losses were not simply due to the length of small bowel resected, but also to factors such as prolapse, stenosis, and adhesions.

Seventeen patients had a definitive procedure. The types of procedures and results are shown in Table 4. There were 4 late deaths (24%). Of these, 1 patient had an ileo-anal anastomosis (Swenson) at 2 days of age followed by a wound infection with signs of peritonitis that responded to conservative treatment. The infant was commenced on oral fluids but had severe diarrhoea with fluid and electrolyte problems. Carmine dye appeared in the stool 10 min after oral intake. After 3 months of intermittent parenteral feeding the infant developed a fatal *Staphylococcus aureus* pneumonia.

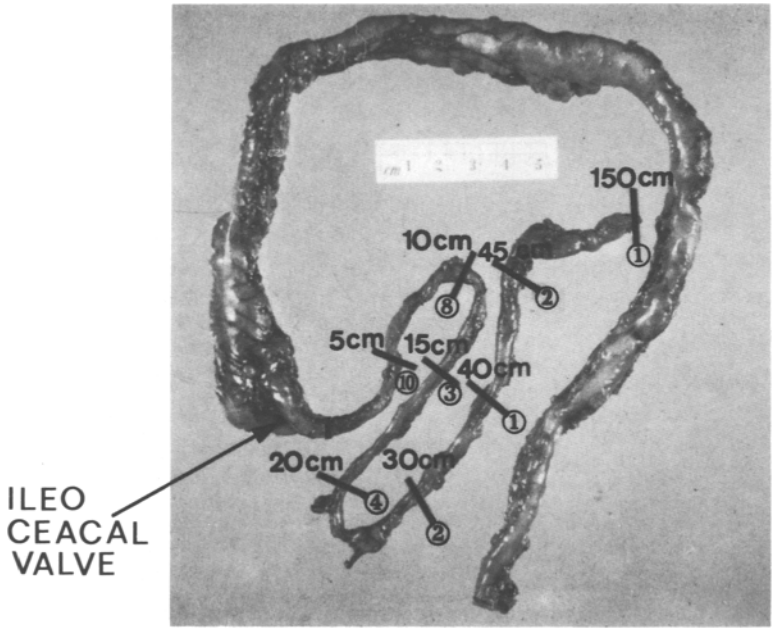

Fig. 3. Site of transition in 31 of the 32 patients with total colonic aganglionosis (site not accurately stated in the other patient). ○ = number of patients

Table 4. Definitive operative procedures and complications in 17 patients with TCA

Procedure	No.	Age	Deaths	Complications				Current status		Average follow-up
				Early		Late				
Swenson	6	Under 6 months: 2 18 months: 4	2	Wound infection Obstruction Abscess	3 1 1	Night soiling until 4, 6, 11 years	3	Normal continence Malabsorption	4 1	15 years
Duhamel[a]	7	9–30 months	2	Wound infection Incisional hernia Stricture Adhesions Obstruction	2 1 1 1 2	Night soiling until 6 years Anterior pouch Abdominal pains Fatal enterocolitis	1 1 1 1	Normal continence Nocturnal incontinence (age 8½) Lost to follow-up	3 1 1	8 years
Soave	4	13–24 months	0	Stricture Obstruction Neo-ileostomy	1 2 1	Abscess ovarian perianal	1 1	Normal continence Nocturnal incontinence (age 8) Malabsorption	3 1 2	5 years

[a] Includes one patient who had a "long" Martin procedure

Another patient had a Swenson ileo-anal anastomosis at 4 months with wound breakdown 14 days after operation, but was subsequently well, requiring only one anal dilatation. Four months after operation the child developed fever and peritonitis and died. Post-mortem examination showed an abscess between loops of ileum about 8 cm from the ileo-anal anastomosis with signs of peritonitis. Therefore, both patients who had had their definitive operation before 6 months of age died. The third death was due to enterocolitis 3 years after a Duhamel operation. The patient had had 2 previous episodes of enterocolitis, 1 month and 11 months after operation, but had subsequently been well after anal dilatation, with 1–2 bowel actions per day, and was continent. The patient had a short history of vomiting with diarrhoea and was misdiagnosed as gastroenteritis. The fourth death occurred some months following a Duhamel operation, but necropsy indicated that the cause of death was neither Hirschsprung's disease nor its treatment.

Complications after the definitive operative procedure included 12 patients with severe excoriation of the buttocks, 5 wound infections, 2 abscesses (1 paracolic, 1 later ovarian), 1 adhesive bowel obstruction and 2 strictures. Five patients had obstructive episodes and 2 of these required additional defunctioning ileostomies which were subsequently closed after anal dilatation. Two patients had sphincterotomy and 1 required the occasional passage of a rectal tube. In the 13 operative survivors, these early obstructive complications settled with time.

Follow-up has been up to 26 years. Day continence was only slightly delayed in 10 patients with achievement of continence by the age of 4. The other 4 patients became continent at ages 5, 6, 8, and 8½ years, respectively. Nocturnal continence was achieved much later (Table 4), and even at 25 years, 1 patient still has to go to the toilet once a night and to avoid certain foods. There were no disorders of urinary continence or sexual function. Four patients had lactose intolerance, which resolved, and 3 had malabsorption and vitamin B_{12} deficiency although they had not had large segments of ileum resected). Weights and heights were available for 10 patients: 6 are on or above the 50th percentile, 1 is on the 10th percentile, and 3 are on or just below the 3rd percentile.

Discussion

The first clinical description of TCA was probably by Porter and Weeks in 1915 [17], although there was no histological confirmation. The first histologically documented patient was described by Zuelzer and Wilson in 1948 [42] and the first successful definitive operation performed by Sandegard in 1953 [34]. The early clinical reports highlighted a mortality of 65%–95% [2, 13, 14, 40], but more recent series suggest improvements in treatment with mortality rates of 0%–28% [1, 6, 24].

Experience in this series corresponds to that of others. The first patient was diagnosed in 1954 and the first survivor appeared in 1957. The mor-

tality was 82% in the 1st decade of this study, 50% in the 2nd, and 9% in the last (Table 1). Of the 15 deaths in the series, 8 were primarily due to delays in diagnosis and control of sepsis (1 was thought to be secondary to congenital asplenia), 1 to prolonged ileostomy dysfunction, and 4 occurred after the definitive operation (Table 2). As in other series, post-operative enterocolitis can be a later fatal complication [6, 22]: it occurred in 1 patient 3 years after a Duhamel procedure. Two other infants who died post-operatively had had an early Swenson procedure (operation at 2 days in one patient and 4 months in another). Swenson [39] reported a high mortality (28%) in infants operated on in the first 6 months of life and advised against early operation. More recently, however, definitive operations of the pull-through type have been performed without complication in neonates with rectosigmoid aganglionosis [35], this procedure has not yet been reported or recommended for infants with TCA. The fourth death occurred after a Duhamel procedure, but was due to unrelated causes. The improvements during the course of this series were due to increased awareness, the use of barium contrast studies, rectal suction biopsy and intra-operative frozen section biopsies, control of sepsis, and nutritional support, with the different operative techniques playing a limited role (Table 4). We have had no experience with manometric studies, but these must be interpreted with caution in excluding TCA [10].

The sex ratios, birth weights, gestations, and ages at presentation were similar to those reported in most series [2, 19, 22, 36, 40]. To get a more accurate idea of the sex ratio, as each series had a small number of cases and chance distortions may arise, the cases in this series were added to others in the literature [1, 2, 6, 9, 13, 14, 19, 22–25, 36, 38, 40]. There were a total of 385 cases and the male: female ratio was 1.8 : 1.0. In patients with more proximal small bowel aganglionosis, the sex incidence was equal or even reversed [19]. While 5 patients (15%) had a positive family history, in only 1 case did the relative also suffer from TCA: this was a father- son relationship, and as far as we can ascertain this had not been described previously. Other reported affected offspring from patients with TCA have had recto-sigmoid aganglionosis: in all, 5 offspring from parents with TCA have been described and 3 of these have had aganglionosis (2/4 males; 1/1 female). The genetic risk for offspring cannot be quantified by these few cases; if the trend continues, however, the risks would be high. The inheritance probably differs according to the length of bowel affected; therefore, a classification specifying the extent of bowel involvement (rectal, sigmoid, long colonic, total colonic, and small bowel aganglionosis [19] would be more meaningful than the terms "short"- and "long"-segment that are often used in the genetic literature [7, 31]. The interplay between genetic and environmental factors is interesting, with the importance of local environmental factors being illustrated by the reports of single monozygotic twins being affected [27 (and Mares AJ, personal communication)]. In other situations genetic factors are clearly predominant, as illustrated by family trees with high transmission to subsequent generations [8]. In addition to aganglionosis, there was a high incidence of congenital deafness and to a lesser extent Waardenburg's syndrome [8]. This suggests a generalised neural crest disorder similar to that found in mice that developed aganglionosis as an autosomal recessive trait [41]. The concept of a"neurocristopathy" is further supported by the occasional association of Hirschsprung's disease with neuroblastoma, as occurred in the sibling of one of our patients and as previously reported [15]. In addition, the neural crest probably plays a part in cardiac and splenic development [21, 26], so that the patient with associated transposition of the great vessels and asplenia would fit into this concept of a neural crest disorder. This interaction of environmental and genetic factors in Hirschsprung's disease indicates that the study of the disorder may aid in gaining a clearer understanding of embryonic development.

The presenting symptoms were vomiting and distension in 90% of cases; in 40% of those documented there was passage of meconium in the first 24 hours, a situation noted in other series [6]. On occasion, the diagnosis of TCA has been delayed until adolescence or adulthood [17, 28, 37] even though symptomatology had dated back to the neonatal period. In this series, 3 infants had no documented symptoms in the neonatal period, with the oldest presenting at 4 months of age. As this was early in the series, subtle symptoms may have been missed. With improving documentation over the last 12 years, all patients have had some symptoms in the first 48 hours and have been diagnosed by 2 weeks of age. However, even with increased awareness there have been recent errors of diagnosis, such as meconium ileus from the barium enema and a misdiagnosis of complicated meconium ileus and necrotising enterocolitis with perforation of the transverse colon at laparotomy. In these latter 2 patients, the routine histological examination of tissue from the stoma revealed the true nature of their disease.

Perforation of the aganglionic bowel occurred in 6 patients: it occurred pre-operatively in 4, with the appendix being the probable source in 1, the aganglionic terminal ileum in another, and the transverse colon in 2. The colonic biopsy site perforated in 2 cases (1 also previously mentioned), and in the last patient the aganglionic colon left in situ after the definitive procedure formed an appendicular-vesical fistula. Neonatal perforation of the appendix has been particularly noted for its association with TCA [1, 16, 23, 38], as have the dangers of leaving aganglionic bowel in situ after the definitive procedure [9].

While the plain abdominal X-ray usually suggested a low bowel obstruction (90%), the barium enema was less diagnostic, there usually being no pathognomonic features [1, 16, 23, 38]. On some occasions there were suggestive features such as shortening of the bowel, irregularity of the mucosa, and delayed evacuation; on other occasions the study was essentially normal; and in some situations the findings were suggestive of meconium ileus (microcolon) or meconium plug. The main clinical value of the barium enema is therefore to rule out other causes of neonatal bowel obstruction, the important point being that a normal (or suspicious) barium study in the presence of abdominal X-ray findings of low bowel obstruction is an indication for rectal suction biopsy.

Ileostomy dysfunction was common, with a 20% prolapse rate and a 25% rate of persistent excessive losses. Ileostomy stenosis and adhesions were the main causes of excessive losses rather than a "short bowel" situation, however, as many of these losses stabilised or improved after refashioning of the ileostomy.

The operative procedure of choice remains a controversial topic. Good results have been reported using the Swenson procedure [1, 39], even when 4–6 cm of rectum are retained [36]. The Duhamel procedure has given good results [1, 6, 9], as have the Rehbein [30], Soave [22], and Martin modifications [5, 24, 25]. In this series the most common procedures used were those of Swenson (6), Duhamel (7), and Soave (4), following the historical trends. The main operation now being performed is the Soave procedure. There was a high incidence (50%) of early obstructive symptoms and persistence of nocturnal faecal incontinence until the age of 4–11 (43%). Six of 12 patients with long-term follow-up were completely well. One had a stricture responding to anal dilatation and 2 (14%) had nocturnal incontinence at the age of 8 years. Three (1 Duhamel, 2 Soave) had intermittent mild obstructive symptoms that responded to naso-gastric suction, intravenous therapy, and rectal tubes. Two had severe obstructive symptoms requiring refasioning of an ileostomy, vigorous dilatations and later closure. Three patients were on or below the 3rd percentile and had features of malabsorption requiring vitamin B_{12} supplementation. This series of 17 operated cases is small, but there was no obviously superior operative technique (Table 4).

The management practice in this series was to wait until bowel motions became semi-solid and then perform one of the standard pull-through procedures. This occurred in most of the patients, but it took up to 24 months for the fluid stools to become less frequent. When the small bowel had demonstrated its absorptive ability, there was little to be gained by leaving aganglionic colon in place for its additional absorptive capacity. If the motions remained fluid after 24 months, a Martin modification of the Duhamel procedure was performed [24]. The results reported by Martin [24], show a very low morbidity with no complications such as buttock excoriation, so while there is no benefit as far as mortality is concerned this procedure or similar variations may aid in lessening morbidity. On the other hand, there are some reports of a higher complication rate after the Martin modification, with enterocolitis being a particular problem [6]. At the moment, there is no perfect surgical procedure that results in attainment of full physiological function. This deficiency is highlighted by the recent publication of other modifications [3, 20, 25, 29, 33]. The improvement of treatment will partly depend on a better understanding of normal peristaltic mechanisms [18] and sphincteric function [12] and further detailed study of the aberrant physiology in patients with TCA.

Conclusion

A great deal of our experience with TCA is now of historical interest; however, it is well to recall the problems which preceded the era of intravenous nutritional support for babies and take note of the changes in morbidity and mortality consequent upon the introduction of new and broad-spectrum antibiotics for the prevention and control of sepsis. There can be little doubt that many of the early deaths in this series would not occur today. Although the period under review commenced but a few years after the first definitive operation for Hirschsprung's disease, and thus covers almost the entire period of the modern his-

tory of this disease, there is still no uniformity of opinion regarding the best operation for TCA and reports regarding many new operative modifications continue to appear in the literature. Quite sophisticated methods of diagnosis are now available, but these will always remain secondary to clinical judgement. TCA epitomises the need for diagnostic acumen as a preliminary to a planned programme of treatment designed to restore the patient to a state of normal health.

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