Total colonic Hirschsprung’s disease: a 28-year experience

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Abstract

Purpose: The aim of this study was to review outcomes after surgical treatment of total colonic Hirschsprung’s disease (TCH).

Methods: Twenty-five records of patients with TCH treated between 1974 and 2002 were reviewed. Follow-up data were collected using a standardized questionnaire. Objective functional outcome was assessed using a scoring system.

Results: Twenty patients had aganglionosis of the colon and distal ileum, 5 of whom had a more extensive condition. One of these 5 patients underwent an endorectal pull-through (ERPT), 1 underwent intestinal transplantation, and 3 died. Four of the remaining 20 patients underwent a primary ERPT, 16 received a stoma as neonates followed by ERPT in 12, and a Martin-Duhamel procedure or Swenson’s operation in 3 (median age, 10.5 months); 1 remains with an ostomy. Postoperative complications included enterocolitis (55%), anal stricture (25%), and perineal excoriation (20%). Mean follow-up were 17.5 years (± 11.1 years). Eighty-nine percent were free of recurrent enterocolitis. Frequency of bowel movements is 1 to 5 per day in 82% of the patients, 18% have 6 or more bowel movements per day. Occasional soiling is noted in 40% (one third of those requiring nighttime diapers). Overall functional outcome was good in 83%. Those patients with the longest follow-up periods had the best stooling scores (P = .04).

Conclusions: Surgical treatment of TCH is associated with a number of complications including recurrent enterocolitis and anal strictures. Long-term outcome is quite favorable.

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1. Methods

Records of 25 patients who were diagnosed and treated for TCH at the Section of Pediatric Surgery of the CS Mott...
Children’s Hospital, University of Michigan, between 1974 and 2002 were reviewed. Total colonic Hirschsprung’s disease was defined as aganglionosis of the entire colon with or without extension into the small intestine. Case patients were analyzed for their descriptive data (sex, age at operations, preoperative, perioperative, and postoperative management, and complications). Follow-up data (if >6 months postoperative) were collected using a standardized telephone questionnaire as outlined in Table 1. This questionnaire allows one to obtain an accurate history of continence as well as the stooling pattern of a patient. Objective functional outcome was assessed using a scoring system (Table 1). If a patient required an ileostomy, he or she was automatically assigned a poor outcome. Data are given as the median with range or mean ± SD. Statistics used multivariate linear regression analysis. The study was reviewed and fully approved by the respective institutional review boards (IRB No. 2000-0450).

2. Results

2.1. Descriptive data

Over a period of 28 years, 12 boys and 13 girls were treated for TCH. In 23 of these 25 patients, initial presentation was in the neonatal period (median age, 3 days; range, 2-45 days). Eighty-three percent of all cases presented with neonatal bowel obstruction, and one patient presented with neonatal enterocolitis. The 2 children who presented with initial symptoms after the neonatal period had intractable constipation. Median age at diagnosis of TCH was 10 days (range, 3-105 days). Twenty patients had Hirschsprung’s disease involving the entire colon, 12 of them included the distal ileum and 5 had either jejunal involvement [2] or total intestinal aganglionosis [3]. Four of the 25 patients underwent a primary endorectal pull-through (ERPT) in the neonatal period, whereby a total colectomy (including the distally involved ileum) and a primary ileoanal anastomosis were performed (median age, 12 days; range, 12-42 days). The remaining 21 patients received a small bowel stoma as neonates (median age, 18 days; range, 3-77 days), a colectomy was performed at the same time in 60% of the patients; none of the babies with extensive aganglionosis had a bowel resection at that time. In 16 of those 21 patients with primary ileostomy, the pull-through was subsequently performed at a median age of 1 year (range, 0.8-15 years), including one patient with aganglionosis reaching the jejunum. In 84% of those patients, an ERPT was performed, 3 had a Martin or Duhamel procedure or Swenson’s operation. In 88% of those children who initially had an ostomy, the ileostomy was closed at the time of the pull-through; 2 underwent ileostomy closure at a later date. One patient with multiple other congenital anomalies never underwent a pull-through and still has a stoma to facilitate care. One baby with total intestinal aganglionosis underwent an intestinal transplantation at the age of 15 months without previous ostomy (6 years postoperatively, the patient was with an ileostomy in place and showed no signs of transplant rejection). The only deaths in our series involved 3 patients with total aganglionosis who died within the first 6 months of life because of severe sepsis. This reflects a preoperative mortality rate of 13% including all patients. No postoperative mortality occurred.

2.2. Surgical complications

2.2.1. Pre–pull-through period

Five of the 25 patients (2 of whom had undergone a previous colectomy) went through one or more bouts of HAEC; 4 of the 5 infants with HAEC were successfully treated with conservative management and one patient had a colectomy performed at the time of the episode of HAEC (Table 2).
2.2.2. Post–pull-through complications

The most common complications were HAEC (55%), anal strictures (25%), wound infections (20%), and perineal excoriation (20%; Table 2). The most prevalent complication was HAEC. Although the numbers are clearly small, no correlation was found between HAEC and the type of pull-through ($P = .53$; $\beta$ coefficient $= .188$; 95% CI, $-1.361-2.236$) when comparing ERPT to other types of pull-through. Anal strictures requiring anal dilatations were also shown not to be related to the type of pull-through ($P = .99$; $\beta$ coefficient $= -0.003$; 95% CI, $-1.470-1.458$); perineal excoriation also showed no correlation ($P = .18$; $\beta$ coefficient $= -.458$; 95% CI, $-3.046-0.651$). Two patients experienced incorrect intraoperative leveling of the pulled-through bowel and subsequently needed an ileostomy (31 and 33 months post–pull-through). In these patients, both frozen and permanent sections from biopsies at the time of the pull-through were positive for ganglion cells.

2.3. Long-term follow-up

Mean follow-up of definitively treated patients was 17.5 years ($\pm 11.1$ years; range, 1.2-30 years) and involved 18 patients (Table 3). At the last follow-up, 16 of these 18 children who had undergone a pull-through (89%) have had no recurrent bouts of HAEC. Mean number of bowel movements (BM) was 4 $\pm 2$ per day (1-10/d). Three patients had more than 6 BM per day (follow-up time of 47, 130, and 304 months, respectively); none stated that he or she was socially impaired and all of them showed a normal urgency period. One had occasional soiling at night. Of all followed up patients, loose stool consistency was seen in 53%; 35% had normal firmness of their feces and 2 patients (12%) had liquid stools (304 and 202 months post–pull-through). Occasional soiling was noted in 38% of the patients, with one third of those partially incontinent patients requiring diapers at night. Urgency was felt to be normal in all patients. Long-term use of metronidazole as a prophylactic measure to prevent recurrent HAEC was needed in 3 patients; however, all of those patients were in the early years of post–pull-through (follow-up time, 14-135 months). Attapulgite or loperamide was also required in 3 patients, 2 of whom had to take the regimen in combination with metronidazole. Diet was normal in all patients; no restrictions have been made in any of them. A stooling score was applied to patients who were older than 3 years. Overall functional outcome of patients was good (score of 11-16) in 83% (15 patients) of all patients who had undergone a pull-through. One patient (6%) showed a fair outcome (score, 6-10; because of soiling, high frequency of BM, and recurrent bouts of HAEC), and the 2 patients with incorrect leveling of the pull-through obviously had a poor outcome (11%; follow-up time, 52 and 36 months, respectively). Mean score of functional outcome was 12.1 $\pm$ 4.0. The type of pull-through had no correlation with the overall functional outcome ($P = .26$; $\beta$ coefficient $= .353$; 95% CI, $-0.482-1.631$). Linear regression analysis showed that those patients with the greatest follow-up periods had the best stooling scores ($P = .04$; $\beta$ coefficient $= .64$; 95% CI, 0.007-0.033).

### Table 3 Functional results of follow-up

<table>
<thead>
<tr>
<th>Results of follow-up (n = 18)</th>
<th>Outcome score $\pm$ SD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Recurrent abdominal distension</td>
<td>1.8 $\pm$ 0.4 89% none</td>
</tr>
<tr>
<td>Frequency of defeaction</td>
<td>1.2 $\pm$ 0.7 82% 1-5/d</td>
</tr>
<tr>
<td>Stool consistency</td>
<td>1.4 $\pm$ 0.6 53% loose</td>
</tr>
<tr>
<td>Soiling</td>
<td>1.6 $\pm$ 0.5 38% occasionally</td>
</tr>
<tr>
<td>Urgency period</td>
<td>2.0 $\pm$ 0.0 100% normal</td>
</tr>
<tr>
<td>Diapers required</td>
<td>1.9 $\pm$ 0.3 88% none</td>
</tr>
<tr>
<td>Long-term use of medication</td>
<td>1.5 $\pm$ 0.8 76% none</td>
</tr>
<tr>
<td>Diet</td>
<td>2.0 $\pm$ 0.0 100% normal</td>
</tr>
<tr>
<td>Overall outcome</td>
<td>12.1 $\pm$ 4.0 83% good</td>
</tr>
</tbody>
</table>

Note the good overall long-term outcome in 83% of patients with TCH.

3. Discussion

Total colonic aganglionosis (TCH) is a severe form of Hirschsprung’s disease. It is most commonly recognized in the neonatal period [4], as was seen in our series. The overall TCH mortality rate in the literature has been reported to be as high as 54% [11]. In recent decades, however, the expected survival for TCH has improved dramatically because of increased awareness and knowledge of the disease and its complications. In our series, the preoperative mortality rate was 13%; all were in patients with total intestinal aganglionosis who were not surgical candidates. Postoperative mortality rate was zero. This may well be because of the extreme awareness of HAEC in our institution, with early recognition and prompt initiation of intensive medical treatment [1,12,13]. However, the morbidity associated with TCH remains high. Once the diagnosis of TCH is made, several considerations need to be addressed. First is whether a primary pull-through should be performed vs an ostomy. Second is whether a partial colectomy should be performed at the time of diagnosis of TCH. In our series, 60% of patients had a colectomy at the time of the first exploratory laparotomy with formation of an ileostomy. One potential difficulty with primary definitive surgical management with total colectomy is the necessity of determining the definitive level of aganglionosis. A frozen section of the appendix is almost always diagnostic [1]. To avoid too extensive or inadequate surgery, the most conservative treatment plan in children with suspected TCH is to place an initial diverting ileostomy and await histological results. However, the 2 patients in our series, in whom incorrect leveling of the pull-through occurred, had, in both frozen and permanent sections, ganglion cells seen. One may assume that the subsequently discovered aganglionosis may have been an acquired sequela post–pull-through.
For the definitive repair of TCH, many techniques and their modifications have been suggested. Data on primary ERPT for TCH have not been well described previously. Its clear advantage is a single operation without the potential problems of an ostomy. However, disadvantages include incorrect leveling of an aganglionic segment, inability to adapt to the more rapid transit time, and the potential for perineal excoriation. In our review, 4 of the 25 patients had a primary ERPT performed in the neonatal period: 1 patient experienced no complications, 2 developed anal strictures, 2 had severe perineal excoriation, and 1 child had an ERPT performed at the transition zone, resulting in the need for an ileostomy. Most of our patients were treated with ERPT approximately 1 year after ileostomy. Our data show that the success rate with this latter technique is favorable. Clearly, a complete comparison between types of pull-throughs is difficult, as most patients had undergone an ERPT; however, no correlation with the type of the pull-through and functional outcome was seen. The authors realize that, because of the low numbers of patients who were treated by procedures other than an ERPT, definitive statements about the functional outcome with a particular procedure are difficult. It appears that any of the 3 standard corrective procedures for Hirschsprung’s disease, ERPT, Duhamel operation, and Swenson’s operation may be used to treat TCH [1,3]. In summary, it seems that the essential issues in TCH are not only the surgical management of the disease but also its prompt diagnosis, the management of presenting symptoms, and the treatment of postoperative HAEC. In our series, the most frequent postoperative complication was HAEC, which was in most cases successfully treated with conservative management. It has been shown that increased length of the aganglionic segment is associated with an increased incidence of HAEC [1]. Another frequently occurring postoperative complication was anal stricture. Although in some reports this has been treated with internal sphincterectomy [8], in our series this was managed with repeated anal dilations. Several additional complications occurred in our patients with TCH, 7 of our 18 pulled-through patients required a subsequent operation, ranging in number from 1 to 5; these results are comparable with those seen in other series.

Follow-up of patients with TCH over many years, however, reveals that TCH in the long term has a favorable outcome. Frequency of BM was scored within acceptable limits in 82% of our patients. In cases where soiling was reported, it was mainly noted at nighttime. It was interesting to see that parents and patients scored the overall functional result to be good to excellent in 89% of all our pull-through-treated patients, and objective scores reached a level of good outcome in 83%. In the large-scale review of TCH by Tsuji et al [8], it was shown that anorectal function improves gradually over time, a phenomenon that has also been observed in our study.

The morbidity remains high in patients with TCH. However, mortality should be expected to be quite low, and the older the patients are, the better they do functionally.

References


Discussion

T. Buchmiller-Crair (New York, NY): I would like to ask a question regarding the incidence of enterocolitis, as it seems to be quite high in this patient population. Do you think there is a role for either prophylactic antibiotics or the use of anal dilations in the postoperative period to try to prevent this complication?

B.E. Wildhaber (response): I think the point that we have a high frequency of enterocolitis in our series is well taken. We are very aware of enterocolitis at the University of Michigan and the mildest signs are treated with washouts and with antibiotics. Maybe Dr Coran wants to add more about prophylactic treatment with antibiotics or dilations.

A.G. Coran (response): The incidence of enterocolitis is very high in this group but it is also relatively high in a
series of overall Hirschsprung’s disease of close to 450 patients. Our threshold for putting these patients on prophylactic Flagyl is very, very low. I think mild enterocolitis is something that is missed very often and mistaken for things like viral gastroenteritis. When we see patients with any mild symptoms like that, we immediately start them on Flagyl and keep them on it for a while. We don’t just give them 10 days of it; rather we give them a couple of months sometimes. I think that does reduce the risk of getting grade 3 or grade 4 enterocolitis. If they are sick, we also add washout to it.

J. Langer (Ontario, Canada): You had 4 patients that you did primary pull-throughs on and the rest had stomas, correct?

B.E. Wildhaber (response): Yes.

J. Langer: I am a big proponent of primary pull-through for Hirschsprung’s disease. However, I think in total colonic disease there may be a very good argument made for doing a stoma and waiting for the stool to thicken up, and then doing the pull-through at that point where you may have a much lower risk of the perianal excoriation which is not life-threatening, but it is a major problem for these families. How do you feel about that, and did you have an increased risk of perianal problems in the primary pull-throughs? Are you advocating primary pull-through for these patients?

B.E. Wildhaber (response): I think we cannot draw any conclusion of these 4 patients of those 25 we had, and we saw a spectrum of every complication in those 4 patients. There is one patient with absolutely no complication at all, and there is the other patient with having anal strictures and enterocolitis and perianal excoriation. And there is the third patient which unfortunately received a pull-through in the transition zone. So the spectrum is from 0 to 100. There is no conclusion we can clearly draw with our data.

A.-X. Holtermann (Chicago, IL): I am struck by your high incidence of anal stricture. Is this a procedure-related technical complication, such as ileal pull-through, because I do not think we see that high of a complication rate with the standard colonic pull-through for rectosigmoid Hirschsprung.

B.E. Wildhaber (response): Our analysis showed that there was no relationship to the type of pull-through, either to Duhamel, Martin, or the rectal pull-through. There was also no relation to the length of the aganglionosis segment. I don’t have an answer. It was just seen that a quarter of them, needed dilations under anesthesia, but no sphincterotomy, no further operations were needed.