

Hirschsprung's disease: functional and psychological follow up comparing total colonic and rectosigmoid aganglionosis

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Aims: To compare the long term functional and psychosocial outcomes following surgical treatment for total colonic aganglionosis (TCA) with those in an age and gender matched group of patients with rectosigmoid aganglionosis (RSA).

Methods: Fifteen patients with TCA matched for age and gender with 15 patients with RSA were studied 7-17 years after the definitive operation. The internal and external sphincters were examined using anal endosonography. Functional outcome (faecal continence) was assessed by a surgeon not involved in the patients' care, and by a research psychologist in separate assessment sessions. The behavioural and emotional status of the patients was also assessed.

Results: Based on the surgeon's assessment, 6/15 TCA and 7/15 RSA patients were continent. In comparison, based on the psychological interview, 2/15 TCA and 6/15 RSA patients were continent. The TCA patients reported significantly more behavioural/emotional problems and lower levels of self esteem than the RSA patients. The parent and teacher assessments of psychosocial status revealed no differences between the groups. There was no association between incontinence and psychosocial adjustment in either group. There was no association between the assessments of functional outcome and the endosonographic appearance of the anal sphincters.

Conclusions: The proportion of patients with faecal incontinence 7-17 years after definitive surgery was high in both groups, but no association was found between incontinence and the psychosocial outcome measures. TCA patients perceived themselves as less well adjusted than their matched pairs. Differences between the groups in length of hospitalisation and severity of illness, especially in infancy and early childhood, may account for these differences.

Total colonic aganglionosis (TCA) accounts for 3-12% of children with Hirschsprung's disease.¹ These patients are considered as an important subset because of the associated increased morbidity and mortality.¹⁻³ Although mortality has decreased in recent years,⁴ patients with TCA still present particular difficulties in diagnosis and treatment and tend to undergo multiple procedures.⁵ A higher proportion of these patients experience episodes of enterocolitis after surgery compared with patients with shorter segment disease.¹ Many are at risk for associated complications such as failure to thrive, stomal dysfunction, electrolyte imbalance, and dehydration.¹

Long term functional outcome of patients with TCA has not been systematically investigated.¹⁻⁵ It is generally assumed that the longer the aganglionosis the poorer the anorectal function.^{1,6}

This study aimed to investigate whether long term continence and psychosocial adjustment is affected by the length of aganglionosis by comparing TCA patients with patients with rectosigmoid aganglionosis matched for age and gender.

PATIENTS AND METHODS

Subjects

Patients with biopsy proven total colonic aganglionosis (TCA) Hirschsprung's disease, who had definitive surgery at the Great Ormond Street Hospital for Children, London between 1979 and 1990, were eligible for participation in this study. Patients were excluded if they had undergone definitive surgery at another institution, had multiple congenital abnormalities, known neurological problems and/or developmental delay, had a permanent stoma, and were non-English speaking. During the 11 year study period 54 patients with

TCA Hirschsprung's disease were operated on at our hospital. Twenty cases fulfilled the inclusion criteria for this review. Of these 20 TCA patients, two declined to participate. Additionally, we were unable to trace three patients. Thus the TCA study sample comprised 15 TCA patients (11 boys, four girls), aged 7-18 years (median 12 years). The aganglionosis in these patients involved the entire colon in five, the colon and terminal ileum (<30 cm) in six, more than 30 cm of ileum in three, and the colon and entire ileum in one. The functional and psychological outcomes of the TCA patients were compared with that of a group of patients with aganglionosis confined to the rectosigmoid colon (RSA), matched for age (median 12, range 7-17 years) and gender (11 boys, four girls), who fulfilled the inclusion criteria.

Ethics

The study was approved by the ethics committee at our institution and informed written consent was obtained from patients and parents.

Outcome measures

The study design included a clinical interview and examination carried out by a surgeon (AP) not involved in the child's original surgery. In addition, one investigator (HT) assessed the status of the internal and external sphincter using anal endosonography. Following the clinical assessment, the child and the parents were interviewed separately by a research

Abbreviations: CBCL, Child Behaviour Checklist; RSA, rectosigmoid aganglionosis; TCA, total colonic aganglionosis

Table 1 Functional assessment scoring methods and evaluation of continence

Surgeon's assessment measure (Kelly score) ⁷		Psychologist's assessment measure			
Frequency of staining/smearing					
Always clean	2				
Occasionally stained	1				
Always stained (>3× wk)	0				
Occurrence of accidental defaecation or soiling					
Normal control under all circumstances with no accidents	2				
Occasional escape of faeces or flatus, with occasional major accidents	1				
No control or frequent accidents (>3× wk)	0				
Puborectalis muscle: sphincter squeeze					
Effective, strong squeeze of puborectalis	2				
Weak or partial squeeze of puborectalis	1				
No contraction detectable	0				
Surgeon's evaluation					
Modified Kelly Score excluding strength of puborectalis muscle (Total score 0–4)	TCA number (%)	RSA number (%)	Psychologist's evaluation (Total score 0–4)	TCA number (%)	RSA number (%)
Full control (score 4)	6 (40%)	7 (47%)	Full control (score 4)	2 (13%)	6 (40%)
Score 3	3 (20%)	4 (27%)	Score 3	6 (40%)	3 (20%)
Score ≤2	6 (40%)	4 (27%)	Score ≤2	7 (47%)	6 (40%)

psychologist (LL) blind to the extent of the disease and to the continence score (Kelly score)⁷ derived from the surgeon's clinical assessment (table 1).

Anal endosonography

The definitive operation for Hirschsprung's disease can produce disruption of the sphincters which in turn can lead to faecal incontinence. For this reason an assessment was made of the appearance of the internal and external anal sphincters using anal endosonography. This was performed using a Bruel and Krajer 3535 ultrasound system (Bruel and Krajer, Denmark) with a 10 MHz rotating endoprobe of 17 mm in diameter. With the subject in the supine position the perineum was inspected and the lubricated endoprobe was gently inserted into the anal canal.⁸ On the basis of endosonography the internal and external sphincters were classified as intact or disrupted.

Psychological evaluation

Patients and their parents were interviewed separately by the research psychologist (LL). The format of the semistructured interviews, adapted from those used in a previous study of children with anorectal anomalies,⁹ included detailed questions about the child/adolescent's bowel function, soiling, and degree of faecal incontinence. Based on this information, the psychologist's continence score was obtained. Faecal continence was graded on a scale of 0 to 4, mirroring the soiling and staining/smearing classifications of the Kelly score (table 1).

Standardised questionnaires

The Child Behaviour Checklist (Achenbach), a questionnaire completed by parents (CBCL), the parallel Teacher Report Form, and the Youth Self-Report Form (patients aged 11 years and over) were used. The Achenbach questionnaires¹⁰ are designed to measure competencies and behavioural/emotional problems as seen by parents, teachers, and youth respectively, and make it possible to compare data from different respondents on a common set of problem items and scales. They have been used extensively in child/adolescent mental health research. Reliability and validity are well established.¹¹

The patients completed the Depression Self Report Scale,¹² an 18 item screening measure for affective disorders in childhood and adolescence. They also completed age appropriate measures of self esteem (Harter Self-Perception Profile for Children (SPP, British version) and adolescents).^{13 14}

Statistical analysis

Continuous variables with or without a normal distribution were expressed as mean (SD) or median (range) respectively. Differences between the groups were tested by matched pairs *t* tests or the Wilcoxon matched pairs signed ranks test.

The correlation between continence outcome and psychosocial outcomes was assessed by the Spearman correlation coefficient. Measurement of agreement was with the kappa (κ) statistic.

The three Achenbach behaviour checklists were scored using a computer scoring programme which calculates the raw scores and standardised scores corrected for sex and age (T scores: mean 50, SD 10) for each scale. T scores above the recommended cutoffs, that is, T scores in the so called clinical range¹⁰ (internalising, externalising, and total problems T scores greater than 63 and greater than the 90th centile), provide the strongest possible distinction between children with psychopathology versus children without.

RESULTS

Table 2 summarises the clinical details of the patients studied.

Continence

Two TCA (13%) and four RSA (27%) patients would not agree to a rectal examination. In order to obtain a continence score for the total sample derived from surgeon's clinical assessment, the score for the strength of the puborectalis muscle (0–2) was excluded. Based on this modified score, six (40%) TCA and seven (47%) RSA patients had full bowel control (total score of 4). In contrast, the psychological interview revealed that two (13%) of the TCA patients and six (40%) RSA patients had full bowel control (table 1). There was moderate inter-rater agreement between the two assessment methods (κ 0.49, 95% CI: 0.25 to 0.73).

Within the TCA group, continence was not influenced by the length of the aganglionosis and the extent of small bowel resection. The length of aganglionic bowel did not correlate with either the surgeon's continence score ($r = 0.112$, $p = 0.69$) or the psychologist's continence score ($r = 0.019$, $p = 0.95$).

Six TCA patients required antidiarrhoeal agents to assist bowel control. There was no association between the use of antidiarrhoeal agents and the length of aganglionic bowel ($r = -0.051$, $p = 0.86$). One RSA patient required antidiarrhoeal agents occasionally and one required bowel washouts on alternate days for constipation.

Table 2 Clinical details

	TCA	RSA
Age pull through (months)	9 (2–24)	9 (3–24)
Number of main operations (related to HD only)	4 (2–6)	3 (2–7)
Patients with additional anomalies	6*	0
Length of hospitalisation (days)	127 (26–637)	50 (20–134)†
Patients who required parenteral nutrition (length of treatment, range)	6 (8 days to 10 mth)	0
Number of stools a day	4 (2–12)	2 (1–6)

Continuous variables are expressed as median (range).

*Associated congenital anomalies included limb deformities in three patients, hiatus hernia, absent kidney, and malrotation.

†Difference between the groups, $p < 0.009$.

Table 3 Behavioural and emotional adjustment findings (matched pairs)

	TCA Mean (SD)	RSA Mean (SD)	Mean difference	95% CI	p value
CBCL					
Total problem score	51.2 (11.1)	52.4 (8.6)	-1.2	-9.9 to 7.5	0.71
Internalising score	51.7 (11.6)	51.2 (12.5)	0.5	-9.1 to 9.9	0.92
Externalising score	47.7 (9.8)	50.9 (8.6)	-3.3	-11.5 to 5.0	0.41
TRF					
Total problem score	48.1 (5.9)	51.5 (10.0)	-3.5	-11.4 to 4.5	0.36
Internalising score	49.7 (8.3)	50.1 (12.5)	-0.4	-8.6 to 7.9	0.92
Externalising score	48.0 (7.5)	50.8 (10.9)	-2.8	-13.2 to 7.5	0.56
Academic performance	321 (89)	326 (57)	-5.0	-53.4 to 43.4	0.83
YSR					
Total problem score	52.5 (6.1)	45.8 (9.0)	6.6	0.2 to 13.0	0.04
Internalising score	52.8 (5.5)	48.1 (9.5)	4.7	-1.4 to 10.9	0.12
Externalising score	50.7 (8.1)	45.1 (8.8)	5.5	-1.2 to 12.3	0.09
DSRS* (high scores worse)	8.1 (3.2)	5.6 (4.5)	2.5	-0.4 to 5.3	0.09
Self esteem† (lower scores worse)	2.8 (0.7)	3.4 (0.6)	-0.6	-1.1 to -0.1	0.03

CBCL, Child Behaviour Checklist; TRF, Teacher's Report Form; YSR, Youth Report Form (standardised scores).

*Birleson Self-report Depression scale; †Harter self-perception scales.

Anorectal endosonography

Six TCA (40%) and seven RSA (47%) patients agreed to the investigation. There was no association between the assessments of functional outcome and the endosonographic appearance of the internal sphincter (surgeon: $r = -0.276$, $p = 0.39$; psychologist: $r = -0.054$, $p = 0.87$). One TCA patient and three RSA patients were shown to have a disrupted internal sphincter. Only one patient (TCA) had a disrupted external sphincter.

Psychological assessment

Table 3 summarises the behavioural and psychological findings. Two of the patient's self rating scales revealed significant differences between the matched pairs. TCA patients reported more behavioural and emotional problems and had lower levels of self esteem compared with their matched pairs. There was no association between incontinence and behavioural adjustment, depressed mood, or self esteem in either group. The parents' and teachers' perceptions of the patients did not concur. Although the parent and teacher assessments revealed no overall differences between the groups on the raw or standardised mean scores (table 3), the frequency distribution of the total problem scores showed that, based on the parental assessments, more TCA than RSA patients (4 (27%) v 2 (13%); $p = 0.2$, 95% CI: -0.15 to 0.42) had high total problem scores (scores more than 1 SD above the mean), indicating a greater likelihood of clinical psychiatric problems. Conversely, more RSA patients (3/13, 23%) than TCA patients (1/13, 8%; $p = 0.15$, 95% CI: -0.12 to 0.42) had high total problem scores based on the teachers' report (two pairs were no longer at school). There was no association between incontinence and the parents' and teachers' assessments.

DISCUSSION

The main purpose of this study was to examine in detail long term functional and psychosocial outcome in patients with total colonic aganglionosis, and to compare their outcomes with an age and gender matched group of patients with an aganglionic segment confined to the rectosigmoid area.

Outcome following surgery for Hirschsprung's disease has generally been evaluated in cohorts of surviving patients, including those with multiple congenital abnormalities such as Down's syndrome, treated in single centres or large single surgeon series. The majority have reported on functional outcome in relation to surgical techniques and complications following corrective surgery. A few recent studies have reported on psychosocial and functional outcome.¹⁵⁻¹⁸ However, outcome measures were often based on replies to postal questionnaires¹⁹⁻²¹ which restrict the investigator's ability for probing symptoms and may be sensitive to the respondent's educational level. Similarly, assessing long term outcome of TCA patients is hampered by the lack of objective reporting and the methods used to assess continence.

The design of our study differs from previous reports. First, we have restricted our sample to two specific types of Hirschsprung's disease, patients with biopsy proven total colonic aganglionosis (TCA) and those with an aganglionic segment confined to the rectosigmoid area (RSA). Second, the patients were carefully matched for age and gender. Third, we report only on patients who agreed to a clinical as well as psychosocial assessment. Fourth, patients were assessed 7–17 years after definitive surgery.

A problem that besets all studies which aim to measure faecal continence is how to obtain full disclosure from patients or

their parents. Various scoring methods have been developed and the Kelly score⁷ has been used extensively. It requires the patient to be physically present and necessitates a rectal examination. However, in our study, patients were reluctant to allow a rectal examination.

The discrepancy between the surgeon's and the psychologist's evaluation of continence may reflect the fact that compared with the surgical assessment, a psychosocial interview provides opportunities to inquire more deeply into difficulties with bowel control. Moreover, interview data²² have revealed that patients are reluctant to admit to bowel control problems when questioned by surgeons, as this may result in further investigations or surgical procedures.

This study shows that long term incontinence is common in patients with Hirschsprung's disease regardless of the extent of aganglionosis. These findings are in line with recent studies¹⁸⁻²¹ which have reported much higher than expected levels of incontinence among patients with Hirschsprung's disease. Despite the high proportion of incontinent patients, we found no association between incontinence and psychosocial adjustment based on either the patients' self report measures or the parents' and teachers' assessments. This replicates the findings from a previous study of a large sample of children/adolescents with anorectal malformations which showed that, although there were relatively high levels of maladjustment among the children/adolescents, those who were incontinent were no less well adjusted than those with bowel control.^{9,23} Others such as Diseth and Emblem²⁴ and Catto-Smith and colleagues¹⁸ reported no association between incontinence and the CBCL (parents' perception of behavioural adjustment). It is possible that the majority of children with congenital abnormalities of the bowel, together with their families, learn over time to cope with chronic physical health problems such as faecal incontinence. Mediating factors such as the parent-child relationship, the parental relationship, and the family's socioeconomic circumstances may have an equally important influence on the child's emotional development and adjustment.

The psychosocial self report questionnaires completed by the patients showed that the TCA patients had higher (or worse) scores on all four measures of outcome. Their overall behavioural adjustment score was significantly higher than that of their matched (RSA) pairs, and they also reported significantly lower levels of self esteem. Some support for these findings come from the parents' assessments since the parents rated a higher proportion of the TCA patients (TCA: 27%; RSA: 13%) as having scores indicative of psychopathology. Forty per cent of TCA patients had associated anomalies and/or required parenteral nutrition. In addition, they were hospitalised for longer periods than RSA patients; this risk factor^{25,26} may have contributed to their less than optimal perceptions of psychological wellbeing when compared with their matched pairs.

A limitation of this study is the sample size. This was unavoidable as TCA accounts for only a small proportion of patients with Hirschsprung's disease. Additionally, as we were interested in assessing functional outcome, we had to exclude patients with a stoma and those with developmental delay.

In conclusion, faecal incontinence affected a high proportion of the patients in this study, regardless of the extent of the disease. Based on the patients' self ratings, the TCA patients were less well adjusted than their matched RSA pairs. We found no association between incontinence and behavioural and emotional adjustment. Compared with the surgeon's evaluation, the psychosocial interview revealed a higher proportion of incontinent patients among the TCA patients, compared with the surgeon's evaluation. This suggests that postoperative functional problems may be under reported, especially to medical staff, and highlights the difficulties of obtaining an objective measure of anorectal function.

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