Pediatr Surg Int (2006) 22: 319–325
DOI 10.1007/s00383-006-1651-6

ORIGINAL ARTICLE

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An example of psychological adjustment in chronic illness: Hirschsprung's disease

Accepted: 23 January 2006 / Published online: 17 February 2006

Abstract The aim of this study was to investigate the outcomes after definitive surgical correction for children with Hirschsprung's disease (HD) and the psychosocial impact of HD on the child and family. The total sample comprised 72 children with HD along with their families. The development of a condition-specific questionnaire measured the functional and psychosocial outcomes for children with HD with parental perception of their child's condition. Psychiatric measures were also examined to assess psychiatric morbidity. The greatest functional problem after definitive surgery for HD was faecal soiling (76%). The principle findings of the study were that (1) HD did not have a significant impact on the child's rate of psychiatric morbidity and levels of hopefulness in comparison to the normal population, (2) surgical and psychosocial functioning improved with increasing age and, (3) families remain troubled about their future with HD and dealing with psychosocial difficulties related to the condition (such as distress because of faecal soiling). Specifically, faecal soiling was found to be physically, emotionally and psychosocially distressing complication. Bowel functioning and psychosocial distress improves with increasing age and parental and medical professional support. Despite the significant impairment of faecal continence, we found that children/young adults with HD have minimal psychiatric morbidity, yet experience condition-specific psychosocial problems (e.g. embarrassment and distress/discomfort). HD does not increase the rate of clinical psychiatric morbidity in children and families with HD, but does determine the context of their daily distress and concern.

Keywords Hirschsprung's disease · Faecal soiling · Bowel function · Psychosocial outcomes · Psychiatric morbidity

Abbreviation HD: Hirschsprung's disease

Introduction

A chronic condition is defined as lasting longer than 3 months, or hospitalisation of 1 month per year [1, 2]. As one would expect, when surgical treatment is carried out on the paediatric patient attention is focused on the immediate outcome. While surgery treats the acute symptoms of bowel obstruction and vomiting in Hirschsprung's disease (HD), ongoing bowel functioning remains a problem.

HD is defined by the congenital lack of ganglion cells in the distal bowel with variable degree of proximal extension [3–6]. It has been recognised for more than 200 years, yet it has only been within the past 40 years with advances in surgical correction and an understanding of the embryogenesis that a more in-depth understanding has developed. With various surgical advances for HD today, the post-operative results after definitive treatment for children with HD generally are satisfactory. However, HD can result in a lifelong illness as some children with HD continue to have persistent bowel dysfunction in their everyday life [5–9].

The literature has primarily concentrated on the functional outcomes for children with HD with little or if any focus on the psychosocial impact HD has on both the child and family [10–13]. Faecal soiling has the greatest impact on the overall psychosocial functioning of children with HD [9, 14, 15]. This is because soiling is embarrassing and is regarded as shameful and socially unacceptable. An accurate measure of continence is of-

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ten difficult to achieve due to definitional variation and failure to make a clear distinction between occasional soiling and significant soiling. Rarely has both parental and children's perception of the long-term outcomes been included [7].

The aim of this study is to examine the overall outcomes for children who have undergone definitive surgery for HD. We specifically want to address the impact of HD on the child and family and the levels of personal hopefulness for the young adult with HD together with their parent's perspective.

Materials and methods

Approval to conduct this study was obtained from the Ethics Committee of The Children's Hospital at Westmead, Australia. Children who were diagnosed and surgically repaired for HD between 1975 and 2000 were included in this study. The original HD sample comprised 131 children living in Australia, aged between 1 and 24 years of age at the time of follow up. The study was designed so that age groups were divided on the basis of major age groups, such as pre-school and school ages. Such age divisions make it possible to evaluate bowel functioning changes and increase awareness for medical professionals of the psychosocial distress that may result due to HD.

Selection and description of participants

Eligibility criteria excluded children with limited medical history (i.e. incomplete medical records), no contact details or who had died. Out of 131 patients with HD, 48 (37%) families could not be located and two (2%) had died due to HD and associated cardiac defects. Previous studies excluded children with Down's syndrome (also developmental delay) and/or TCA patients to avoid bias outcomes in relation to bowel functioning. These children were not excluded in our study. It is vital to investigate all aspects of HD in order to become aware of potential risk groups. Altogether, 81 (62%) out of a total of 131 patients with HD were initially contacted. Nine (11%) did not wish to participate in this research due to emotional burden, time constraints or language barrier (however, interpreters were provided if needed). The total research cohort therefore comprised 72 children with HD including 82% males and 18% females (mainly born at normal weight) aged between 1 and 24 years (Mean Age 12.41; SD = 8.26) (Table 1). As presented in the literature, the majority of children with HD in our sample were male born at full term, which is consistent with the expected male to female ratio (4:1) [16–18].

Technical information

Biographical details and medical history of the child with HD and family medical history were investigated.

Table 1 Frequency of age groups in HD sample[a]

HD sample	Age groups	*N* (%)	Mean (SD)
Group 1	< 7 years old	30 (41)	2.94 (1.57)
Group 2	7–12 years old	19 (26)	9.11 (2.11)
Group 3	13–18 years old	17 (24)	15.41 (2.18)
Group 4	> 18 years old	6 (8.0)	22.17 (1.94)
Total		72 (100.0)	12.41 (8.26)

[a]Reasons for age subgroups are discussed in Materials and methods

A questionnaire was specifically constructed to measure bowel function and psychosocial outcomes for children with HD titled "The Hirschsprung's Disease Family Impact Questionnaire". Previously published questionnaires for assessing the outcomes for HD were felt to provide insufficient detail for the purposes of this study [7, 13, 15, 19–21]. They lacked purpose-design and/or definitional clarity for specific complications (such as soiling and constipation) and failed to address the different target groups involved (such as severity groups). In order to develop a comprehensive questionnaire, extensive collaboration was undertaken, with both the Department of Academic Surgery and Psychological Medicine at The Children's Hospital at Westmead. Input was obtained from a combination of surgeons, psychiatrist, clinical psychologist, statisticians and medical researchers. A draft HD family impact questionnaire was written after consultation and trialled with volunteers ($n = 10$) similar to the target subjects and several health professionals (psychiatrist, surgeons, paediatricians and medical researchers). Volunteers were asked to comment on any potential difficulties in filling out the questionnaires, degree of reading difficulty and clarity of the questions. The final version of the HD family impact questionnaire was administered as part of this study.

To assess psychiatric morbidity in children over the age of seven and the child's parent, validated psychiatric measures were included. Self-reported depression was assessed by the children's depression inventory (CDI) [22, 23] in all children over 7 years old. Anxiety was assessed using the state-trait anxiety inventory for children (STAIC) in those < 13 and the state-trait anxiety inventory for adult (STAI) in those 13 and over [24, 25]. Self-concept was assessed by the self-description questionnaire [26,–28] in all children over 7 years old. Hunter opinions personal expectations scale (HOPES) looked at hopefulness [29, 30] which was assessed in children 12 years and over and completed by the child's parent. Both the child with HD (12 years and over) and their parent completed family environmental scale (FES) to assess family functioning [31–33]. Parents completed a child behaviour checklist (CBCL) [34, 35] describing their child's behaviour and emotional adjustment. Additionally, each parent completed a general health questionnaire (GHQ-28) to measure paternal psychological distress/dysfunction [36, 37].

Statistics

Statistical analysis was performed using the statistical package for the social sciences (SPSS) Version 10.0 [38]. Pearson's bivariate correlation using chi-squared analyses was used to examine relationships variables. A Fisher's exact test was used for any table where there were small numbers (less than five expected cases).

Results

Description of sample

There were 72 children with HD aged between 1 and 24 years (Table 1). Sixteen (22%) had HD with an associated anomaly, the most common being Down's syndrome (10%). Rectosigmoid was the most common length of aganglionosis (Table 2). Thirty-one percent of the HD sample had a family psychiatric history (e.g. more than one member having depression, alcoholism, schizophrenia and suicide).

Outcomes for children with HD

About half of the HD sample, were toilet trained (53%), with 76% experiencing long-term faecal soiling (Table 3). Thus, the most frequent functional problems found in our HD sample were faecal soiling (76%), followed by diarrhoea (69%) and constipation (60%) (Table 3). Forty-nine percent of children were aware when having bowel movements with 21% unaware (some of which included children with HD and Down's syndrome) and 25% remaining uncertain (5% who did not answer this question). Seventy-five percent of the young adults with HD and parents were satisfied with their bowel functioning, yet 17% still remain unsatisfied and 4% uncertain (4% did not answer this question).

Table 2 Length of aganglionosis in HD sample

Length of aganglionosis	Cumulative frequency (%)
[a]Colonic transitional	56 (78)
Ultrashort	0 (0.0)
Rectal	13 (18)
Sigmoid	12 (35)
Upper sigmoid	3 (39)
Lower sigmoid	3 (43)
Rectosigmoid	19 (69)
Splenic flexure	1 (71)
Ascending colon	0 (71)
Descending colon	3 (75)
Transverse colon	2 (78)
Total colonic aganglionosis	14 (100)
Jejunal	0(100)
Total	72 (100)

[a]Colonic transitional group include patients without TCA

Table 3 Frequency of bowel dysfunction in HD sample

Bowel complications	$N = 72$ (%)
Number of children toilet trained	38 (53)
Incidence of soiling	29/38 (76)
[a]Classification of soiling	
Minimal	7 (24)
Mild	21 (72)
Severe	1 (3)
Frequency of soiling	
Daily–weekly	19 (66)
Monthly–yearly	4 (14)
Almost never	6 (21)
Time of day soiling occurred	
Day	6 (30)
Night	3 (15)
Both day and night	11 (55)
[b]Incidence of constipation	
Classification of constipation	43 (60)
Difficulty or delay in the passage of stools	12 (28)
Difficulty in passing stools because they are hard or small	2 (5.0)
Pain while passing stools	2 (5.0)
Combination of all three classifications	27 (63)
[b]Incidence of diarrhoea	50 (69)

[a]Minimal = small amount of staining on underpants, mild = mild staining on underpants/some pieces of faeces on underpants and severe = lots of faeces/staining on underpants
[b]Children were able to choose more than one complication if they needed to

Outcomes for children with HD in relation to surgical procedure

There was no statistical difference between the primary pull through and staged pull through in relation to bowel functional outcome. Although not significant, staged pull through procedures had a higher incidence of diarrhoea (74%) in comparison to the primary pull through (58%). Additionally, there are two operative techniques that are used for the treatment HD (whether it is a primary or staged pull through). Children who had the Duhamel technique had significantly ($P = 0.005$) higher incidence of constipation (66%) than children who had the Soave technique (55%).

Outcomes for children with HD in relation to length of aganglionosis

Children with the most severe case of HD (i.e. TCA) did not have a significantly higher surgical complication rate than those without TCA. In fact, our findings indicated significantly higher incidence of complications in patients without TCA (42%) compared to children with TCA (34%). Patients with other shorter segments of aganglionosis presented with more constipation (68% vs. 29%; p = 0.002), diarrhoea (71% vs. 57%; p = 0.04) and more soiling (41% vs. 36%; p = 0.002) in comparison to children with TCA. Children with HD and an associated anomaly (e.g. Downs Syndrome) had a higher

total complication rate (47.5%) than children with HD without an associate anomaly (38%).

Outcomes for Children with HD in relation to Age

Children with HD ≤ 12 years had a significantly higher incidence of constipation ($P = 0.03$, 65%) and soiling ($P = 0.004$, 59%) in comparison to children > 12 years (48, 22%, respectively). Overall bowel functioning (e.g. constipation and faecal soiling) was worse in children ≤ 12 years in comparison to children > 12 years (Fig. 1).

Outcomes for children with HD from a psychosocial perspective

Children ≤ 12 years were psychosocially more distressed (such as embarrassment, distress/discomfort and family difficulties) due to bowel dysfunction, in comparison to children > 12 years (Fig. 1). Bowel dysfunction causing children ≤ 12 years to feel distressed included constipation, diarrhoea, faecal soiling, abdominal pain and flatulence. Children ≤ 12 years missed significantly more school due to bowel dysfunction (51%) compared to children > 12 years (4%) ($P = 0.004$). HD had an impact on what the child was able to consume in terms of food. Children ≤ 12 years with HD avoided particular foods in their diet more (53%) (i.e. in order to improve stooling patterns) compared to children > 12 years (22%) ($P = 0.001$). Foods avoided varied between individuals (e.g. sweets, dairy products, citrus fruits, yeast, carbohydrates and soft drinks were common foods avoided). Half of our young adults with HD (> 12 years) endorsed feelings of depression, confusion and anxiety, and found it difficult to get interested about their future with HD. Yet, 50% of young adults with HD remain confident and 63% hopeful about their future.

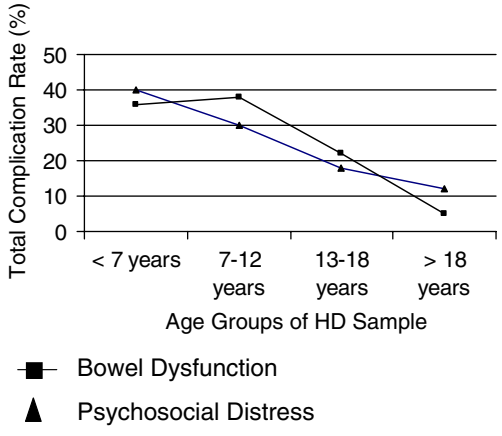

Fig. 1 Total complication rate (%): Includes constipation, faecal soiling, diarrhoea, etc.) calculated using the HD family impact questionnaire

HD had a significant negative impact on family/social functioning. Twenty-eight percent ($n = 20/72$) of the families experienced financial difficulties due to HD (e.g. medication and nappy supplies) (these being significant in families with children ≤ 12 years; $P = 0.02$, 37%) (Table 4). Twenty-five families (out of 72) reported difficulty in public places (such as shopping centres, travelling, beaches, parks and school) due to HD (e.g. faecal soiling) (Table 4). Nine (13%) of the parents reported that HD had a negative impact on their marital relationship due to the daily stressors and associated bowel dysfunction (Table 4). Parents endorsed feelings of depression (32%), confusion (46%), anxiety (56%) and difficulty in getting interested in their child's future with HD (11%). Yet, 58% families remained confident and 71% hopeful about their child's future with HD. Half of the parents in our sample reported family/extended family as the most important support when caring for a child with HD.

Psychiatric morbidity

Overall, 18% children with HD were clinically depressed, 24% with high state anxiety, 56% with high trait anxiety, 4% with behavioural problems and 38% with low self-concept (Table 5). Children with HD > 12 years were more clinically depressed and anxious than children ≤ 12 years. Children ≤ 12 years presented with more interpersonal problems, behavioural problems and high trait anxiety than children > 12 years. Young adults presented with lower levels of personal hopefulness about their future in general (38%) in comparison to their future with HD (54%) (Table 5). Despite ongoing surgical complications found in children with HD (especially faecal soiling) families presented with 79% high cohesion and 68% with low conflict.

Parental morbidity was found to be minimal in our HD sample (17%) with no significant relationship with children's psychiatric morbidity. Parents presented with lower levels of hopefulness about their child's future

Table 4 Correlation between HD impact on family and child's age

Impact on family	≤ 12 years old, $N = 49$ (%)	> 12 years old, $N = 23$ (%)	P	χ^2
Family outings (e.g. party)	13 (27)	5 (22)	0.74	0.11
Work	15(31)	2 (9.0)	0.07	5.24
Family/friend functions	5 (10)	2 (9.0)	0.62	0.96
Financial security	18 (37)	2 (9.0)	0.02	7.54
Siblings	9 (18)	5 (22)	0.47	1.50
Marriage	6 (12)	3 (13)	0.63	0.93
Other impacts	1 (2.0)	2 (9.0)	0.33	2.23

Note: four (5.6%) did not wish to participate in this section of the questionnaire

Table 5 Psychological symptoms in the HD sample

Psychological variables	Age groups (years)	*N* (%)
Clinically depressed (CDI)	7–24	6/34 (17.6)
High state anxiety (STAIC)	8–24	8/34 (23.5)
High trait anxiety (STAIC)	8–24	19/34 (55.9)
Behavioural problems (CBCL)	4–18	2/47 (4.3)
Low self-concept (SDQ)	7–24	13/34 (38.2)
[a]Low level of hopefulness (general) (HOPES)	13–24	9 (37.6)
[b]Low level of hopefulness (with HD) (HOPES)	13–24	13 (54.2)

[a]Level of hope for the future in general
[b]Level of hope for the future with HD

with HD (51%) in comparison to their own future (46%).

Discussion

This study confirms what others have found, that children with HD have significant ongoing difficulties that has an impact on both the child and family's quality of life. Faecal soiling was the most common problem for children after surgical repair for HD (76%). Others have found it was the most frequent and troublesome complication in as many as 32–80% of patients, while others have found reduced frequencies from 5.0 to 30% [9, 15, 17, 20, 39].

Bowel functioning and psychosocial distress improves with increasing age. The literature agrees with our results, suggesting that the bowel function improves over time following surgical correction for HD [8, 15, 17, 21, 39–4142]. Ludman et al. [43] suggested that 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 soiling.

Patients with TCA have generally been considered a subset because of the related increase in morbidity and mortality [44]. The procedure of removing the entire colon from the faecal stream encourages problems with defecation, fluid and electrolyte metabolism [45]. Yet, our study found no significant difference between incidence of bowel dysfunction and length of aganglionosis. Other authors have found long-term faecal incontinence occurring in children with HD regardless of the extent of aganglionosis [7, 8, 13, 39] Our study suggests that other factors could be responsible for ongoing faecal incontinence such as age at which child is followed-up, management and care of the child at home and/or underlying physiological defects which need to be further studied.

In our study, children ≤ 12 years experienced higher levels of embarrassment, distress, discomfort and family difficulties compared to children >12 years. This is generally supported by the literature [9, 14, 46]. Bai [46] found some children with HD to experience moments of embarrassment due to soiling, flatulence and muddy stools resulting in peer relationship problems. However, much of the literature has serious limitations. It fails to differentiate between general psychological symptoms in the child and psychosocial problems specific to HD.

Despite the significant impairment of faecal continence, we found that children and young adults with HD have minimal psychiatric morbidity. Our findings were surprising considering that other studies of children with chronic physical illness have showed twice the expected rate of psychiatric disorders [47–49]. In our study, children ≤ 12 years presented with more interpersonal and behavioural problems than children >12 years. Yet, children >12 years were found to be more clinically depressed and anxious in comparison to children ≤ 12 years. These findings reflect normal developmental changes found in the literature [14, 15, 45]. Yet, it has been found that children and adolescents with HD do not have more psychosocial difficulties than their healthy peers.

This study demonstrated good family functioning with high cohesion and low conflict and low levels of parental psychiatric morbidity. These findings are supported by the literature [50]. In our study, psychosocial distress was specifically due to the daily stressors HD presents to families. Psychosocial distress found in families due to HD included financial burden and difficulty in public places such as shopping centres. Travelling restrictions and restrictions on visiting friends have been suggested as contributing factors to poor family functioning [9]. However, it was suggested that this might appear to improve with increasing age of the child and when independence from his or her parents is established. Other literature has supported such findings also in other chronic conditions (such as epilepsy) [51].

This is the first study to introduce the concept of hopefulness in children with HD. Information on how young adults with HD and their families perceive their future offers greater comprehension of the adjustment required and the impact of HD on the child and family. It was found that some young adults endorse feelings of depression, confusion and anxiety when thinking about their future with HD. Yet collectively with parental support, young adults remain confident and hopeful about their future. Thus, the focus area in future research and clinical management is to counsel these children before their 'distress' transforms into a psychiatric disorder (such as clinical depression). More than half of the parents in the HD sample endorsed feelings of depression, confusion and anxiety about their child's future with HD. Yet, with family support, parents remain confident and hopeful about their child's future with HD.

This study did not include a control group, so it is difficult to compare the results with population's samples or children with other chronic illnesses. However, an attempt was made to accurately describe the severity of physical symptoms in this sample, unlike many other studies, providing a useful window into the day-to-day functioning of a child or young person with HD.

Because children with HD have persistent bowel functioning problems after surgical correction, it is crucial to tell parents about this after surgery, so they have realistic expectations of their child's level of functioning. Counselling should be offered for both the child with HD and their family. Our results indicate that children with HD have good psychiatric morbidity yet experience distress and anxiety specifically due to bowel dysfunction. As this bowel dysfunction appears to be a direct consequence of the disorder, it is important that they are given strategies to cope with the physical problems. Failure to address psychosocial problems from an early age could increase the development of psychiatric morbidity in these children and families with HD. Possible venues for education include seminars or workshops to for health professionals, children and families with HD. Teams treating HD should have early and easy access to psychological consultation.

Management of HD not only involves acute surgical care, but also the need to achieve as close to normal bowel function as possible. We would recommend emphasising the need for long-term regular follow-ups, medication therapy and toilet training strategies. Regular follow-ups will allow physicians to monitor the child's bowel functioning and prevent complications such as faecal soiling from worsening into adulthood. When faecal soiling continues despite active treatment, management strategies to minimise social distress and embarrassment are essential.

In conclusion, it is clear that HD does not inevitably imply that there will be psychosocial problems despite persisting bowel dysfunction. However, when caring for child with HD and their families, doctors need to be aware of the impact of HD on psychosocial well-being for the child and family involved. Living with HD involves not only dealing with the underlying aetiology of the condition, but also the stressors, confusion, hardship, lack of hopefulness and emotional burden it places on the child and family involved.

Acknowledgements We would like to thank Dr Heather Swanston's support for the assisting in the construction of the "HD Family Impact Questionnaire". We would also like to thank all the patients and their families for participating and sharing their experience with us.

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