Chapter 11

Long Term Outcome After Surgery for Anorectal Malformation

Anthony G. Catto-Smith, Misel Trajanovska and Russell Taylor

Additional information is available at the end of the chapter

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1. Introduction

Anorectal malformations (ARM) are a spectrum of congenital caudal end defects affecting the normal development of the anus and rectum that are usually easily detected after birth by physical examination. They are relatively common, occurring in approximately 1 in 5,000 live births, and vary markedly in severity. ARM is frequently associated with other congenital anomalies that may have a much more severe impact on long-term prognosis [1]. ARM are classified into low, intermediate and high subtypes depending on the relationship of the defect to the pelvic floor musculature [2]. A variety of surgical procedures have been developed for their correction, but despite this faecal incontinence is a well-recognized long-term complication that is said to occur in between 10% to 33% or more of patients with ARM [3-6].

Most reviews of outcome after surgical repair of ARM grade continence into broad classifications of "good", "fair" or "poor", based on scoring systems developed by Kelly [7], Templeton [5], Kiesewetter [8] or Wingspread [9]. Faecal incontinence has a significant impact on quality of life in both children and adults [10]. These classification systems have been crucially important in providing statistical information about incontinence but provide little help in understanding how and when incontinence occurs, and its impact at an individual level.

The aims of our study were to define the extent, severity and types of faecal incontinence in children after surgical repair of anorectal malformations, as well as its impact on health care usage, medication, dietary intake, and quality of life. We also wished to relate these outcomes to the level of abnormality, whether "high", "intermediate" or "low".

2. Methods

2.1. Ethical approval

Ethical approval for this study was obtained from the Ethics in Human Research Committee of the Royal Children's Hospital (Approval number EHRC 21118B)

2.2. Subjects

We planned to recruit all patients who had presented for treatment of anorectal malformations to the Royal Children's Hospital, Melbourne between 1974 and 2001. Subjects were identified through ICD coding of medical records. We included patients coded with the diagnoses of anorectal agenesis, rectal atresia, anal agenesis, anorectal stenosis (1970-80; ICD 751.30-33); colonic atresia and stenosis, imperforate anus, anorectal agenesis, anal agenesis (1981-88; ICD 751.20-23); atresia and stenosis of large intestine, rectum and anal canal (1989-2002; ICD-10-AM, 751.2, q42.1-3, q42.18, q 43.6).

We excluded those who were deceased, or non-English speaking, or permanently resident overseas. Clinical information was obtained from medical records. Patients were sub-divided into either a combined high and intermediate category or a low malformation category based on coding information. We also inspected operative notes and separately confirmed correct classification of the level of malformation using the Wingspread Conference criteria [11]. Most patients had the same classification irrespective of the method used. Discrepancies between the two were resolved after inspection of the notes by a paediatric surgeon (RGT).

Recruitment packs were mailed to the most current address recorded in the hospital records. Each recruitment pack contained a detailed information sheet about the study and an initial consent form to allow further discussion of the project. Upon receipt of the signed consent form, we contacted each subject or parent by phone to provide further detailed information about the project and to schedule a detailed interview to administer a structured questionnaire.

2.3. Questionnaire and diary

The 70-item questionnaire was administered either in person, by phone or by mail. The questionnaire had been modified from previous studies on outcome after surgical repair of Hirschsprung disease [12-14]. The revised questionnaire incorporated more detailed information related to bowel function, including amount and awareness of soiling as well as constipation. Further questions were included which related to physical and social functioning, current provision of health care and general health status. The questionnaire was further modified to enable it to be completed by patients as well as parents.

The revised questionnaire was piloted on 21 patients with Hirschsprung's disease. No significant further modifications were required.

Following the interview, respondents were asked to complete a four-week diary detailing bowel habit, faecal soiling, abdominal pain, enuresis and any medical therapy. This was also based on the diary used for the previous study [12]. Soiling was classified into two groups

("light" and "heavy") using the amount and frequency of episode. "Heavy" soiling was classified as being larger than a streak (able to be scraped from underclothing) and occurring at least once a month. "Light" soiling was classified as being no larger than a streak (unable to be scraped from underclothing) and occurring less than once a month. Functional testing, including anorectal manometry and electromyography, was offered [15].

Data from patients who had Down's syndrome was analysed separately.

2.4. Statistics

Statistical analysis was performed using SPSS version 12.0 (Statistical Package for the Social Sciences). Where appropriate, we used t-test, One-Way ANOVA, Chi-square or Fisher's Exact test, Mann-Whitney U Test or Kruskal-Wallis Test. Results were considered significant if $p<0.05$. Data are reported as mean ± standard deviation. Patients who were still in nappies because they were too young to be toilet trained were excluded from some analyses. Missing data are reflected in *n* values.

3. Results

353 patients were identified from ICD coding. We excluded 21 patients for whom we could not obtain permission from the treating surgeon. This was primarily because these surgeons were no longer in practice and had the effect of reducing the number of older patients. A further 34 patients were excluded who were deceased, 2 from overseas, 2 who needed an interpreter and 8 with inadequate contact details. 286 recruitment packs were mailed. Of these, 67 were returned by the post office with "wrong address" and no forwarding details, and no responses were obtained from a further 110 patients. 35 respondents declined or were unable to participate in the study. 74 respondents agreed to participate in the study, of whom 67 were interviewed. Two of the interviewed patients were later excluded because of an incorrect diagnosis. 57 were interviewed over the phone, 7 face to face and 1 via mailed questionnaires. Three patients had Down's syndrome.

The average age of the 62 interviewed patients without Down's was 11.4 ± 6.4yr; 41 were male and 21 female. The average age of the patients who were presumed to be alive but not interviewed was 18.2 ± 8.4yr of whom 428 were male and 275 female. Interviewed patients were younger ($p<0.001$) but gender proportions were similar ($p=0.59$). Four patients who wore nappies were excluded from some analyses. Two wore nappies only overnight (mean age 5.2 ± 1.6yr) and two were too young to be toilet trained, wearing nappies both day and night (mean age 2.4 ± 0.1yr).

12 patients underwent anorectal manometry and electromyography.

Clinical Data (n=62) (Table 1)

There were 28 patients with high/intermediate and 34 with low anorectal malformations. Both groups were similar in age, occupational prestige and family status. Eight patients who had

developed significant faecal incontinence had undergone redo posterior sagittal ano-recto-plasty (PSARP) [16]. Of these, seven had high/intermediate malformations. Patients with high/intermediate malformations were more likely to have co-morbidity.

HD	High/Intermediate (n=28)	Low (n=34)	p	Total
Age (years)	11.8 ± 6.2	11.1 ± 6.7	0.65	11.4 ± 6.4
Gender	21M:7F	20M:14F	0.18	41M:21F
Daniel Occupational Prestige Score	4.4 ± 1.0	4.3 ± 0.9	0.81	4.4 ± 1.0
Single Parent Family n (%) n=62	7 (11.5)	7 (11.5)	0.68	14 (23)
Co-morbidities n (%) n=62	21 (34)	15 (24)	0.01	36 (58)
Subsequent PSARP	7 (87.5)	1 (12.5)	0.02	8 (100)

Table 1. Clinical Data for interviewed patients.

Bowel Habit (n=60) (Table 2)

Patients with a past history of either high/intermediate or low malformations passed a similar number of bowel actions each week (5.5 ± 1.8). Patients with a history of PSARP had more bowel actions each week, although this just failed to reach statistical significance (PSARP 6.6 ± 1.1 vs no PSARP 5.3 ± 1.8, p=0.052). Patients with high/intermediate malformations were more likely to have a looser (liquid or pasty) stool consistency.

		High / Intermediate	Low	p	Total
Bowel actions per week		5.9 ± 1.6	5.2 ± 1.9	0.13	5.5 ± 1.9
Stool consistency n (%)					
	Liquid	3 (10.7)	1 (3.1)		4 (6.7)
	Pasty	11 (39.3)	6 (18.8)		17 (28.3)
	Formed	7 (25)	21 (65.6)		28 (46.7)
	Variable	7 (25)	4 (12.5)	0.01	11 (18.3)

Table 2. Bowel habit data for interviewed patients.

Faecal Continence (n=57) (Table 3)

Impairment in sensation of impending stool was common (38%), as was faecal urgency (74%), difficulty in holding back stool (40%) or controlling flatus (48%) and discriminating stool type (33%).

Patients with high/intermediate malformations were more likely to have difficulty sensing impending stool and holding it back. Younger patients were more likely to experience episodes of faecal urgency (urgency: young 0-11yr 30/36 vs old 12-33yr 12/21, $p<0.05$). Patients were

more likely to have had episodes of faecal soiling during the last year when they also reported problems with sensation, holding back stool, urgency and stool discrimination. (Impaired sensation of impending stool: soiling 17/35 vs no soiling 3/26, $p<0.05$; inability to hold back stool: soiling 22/38 vs no soiling 0/15, $p<0.001$; faecal urgency: soiling 34/42 vs no soiling 8/15, $p<0.05$; or problems with stool discrimination: soiling 14/30 vs no soiling 1/14, $p<0.05$).

		High/Intermediate n (%)	Low n (%)	p	Total n (%)
Sensation impending stool (n=53)	Never/Rarely/Sometimes	15 (62.5)	5 (17.2)		20 (37.7)
	Often/Always	9 (37.5)	24 (82.8)	0.00	33 (62.3)
Faecal urgency (n=57)	Yes	17 (65.4)	25 (80.6)	0.19	42 (73.7)
Faecal urgency – frequency (n=36)	Rarely/Sometimes	9 (69.2)	12 (52.2)		21 (58.3)
	Often/Always	4 (30.8)	11 (47.8)	0.32	15 (41.7)
Ability to hold back stool (n=55)	Never/Rarely/Sometimes	14 (53.8)	8 (27.6)		22 (40)
	Often/Always	12 (46.2)	21 (72.4)	0.05	33 (60)
If able to hold back stool – how long? (n=44)	Seconds	4 (21.0)	2 (8)		6 (13.6)
	Couple minutes	12 (63.2)	16 (64)		28 (63.6)
	Greater than 30 mins	3 (15.8)	7 (28)	0.38	10 (22.7)
Discriminates stool consistencies (n=45)	Never/Sometimes	9 (45)	6 (24)		15 (33.3)
	Always	11 (55)	19 (76)	0.14	30 (66.7)
Stool discrimination type (n=34)	Formed & liquid	0 (-)	1 (5.3)		1 (2.9)
	Formed & gaseous	12 (80)	11 (57.9)		23 (67.7)
	Formed & liquid & gaseous	3 (20)	7 (36.8)	0.36	10 (29.4)
Uncontrolled flatus (n=29)	Never/Rarely/Sometimes	7 (50)	8 (53.3)	0.86	15 (51.7)
	Often / Always	7 (50)	7 (46.7)		14 (48.3)

Table 3. Continence data for interviewed patients.

Most patients (44/60, 73%) had soiling accidents at least once in the past year and this happened "often or always" in 22. 24 soiled only during the day and 20 soiled both during the day and

overnight. There were none who soiled only at night. A higher proportion of females reported soiling episodes (female 19/21 vs male 25/39, $p<0.05$). 43 patients were able to quantify the usual size of soiling episodes that occurred during the daytime. This was medium to large in 26 (60%). Of the 18 who were able to describe the consistency of daytime soiling, 9 (50%) indicated that it was pasty. Eleven had episodes of daytime soiling but were usually unaware that these were occurring (more than 75% of the time). Nocturnal soiling was of pasty consistency in 60% (3/5) patients and medium to large amount in 43% (6/14).

Patients with a past history of high/intermediate malformations were more likely to "often or always" soil during the daytime than those with low malformations (soiling often/always: high/intermediate 15/28 vs low 7/31, $p<0.05$). Despite their revision, those with a PSARP still soiled more frequently (soiling often/always: PSARP 7/8 vs no PSARP 15/51, $p<0.01$). They were also more likely to soil at night (soiling often/always: PSARP 2/7 vs no PSARP 1/48, $p<0.05$).

Most patients could be classified as having either heavy or light soiling based upon frequency and amount of episodes. Nine patients (daytime soiling 1, nocturnal soiling 9) were unable to accurately define the amount or frequency of their soiling episodes. A third (12/33) of patients had "heavy" soiling. "Heavy" soiling was more likely to occur in patients with a high/intermediate malformation ("heavy"soiling: high/intermediate 10/16 vs low 2/17, $p<0.01$). It was also more common in males ("heavy" soiling: male 11/20 vs female 1/13, $p<0.01$) and patients who had undergone a PSARP ("heavy"soiling: PSARP 4/6, no PSARP 8/27, $p<0.05$).

Constipation (n=61) (Table 4)

We defined constipation as fewer than 3 stools per week or straining at stool for more than 25% of the time. Only 13% reported constipation occurring more than once a week. However, episodic constipation did occur. Twelve reported infrequent stools, 34 had hard stools and 39 had difficulty evacuating stools during the past year.

Patients with a low malformation were more likely to have had episodes of infrequent stooling during the past year ($p<0.01$).

Diarrhoea and rectal prolapse (n=57) (Table 5)

12% (7/57) of patients reported that episodes of diarrhoea occurred more than once a week. Patients with high/intermediate malformations were more likely to experience frequent episodes of diarrhoea.

Three patients (6% of 52) had constant anal excoriation and one had a rectal prolapse during the past year. Patients with a high/intermediate malformation were more likely to have perianal excoriation. Patients who had undergone a PSARP were also more likely to have perianal excoriation (excoriation: PSARP 3/8 vs no PSARP 6/44, $p<0.05$).

70% (42/60) of patients had episodic abdominal pain lasting more than two minutes. Of these, 10 reported abdominal pain 1 to 3 times per week and 9 reported pain 1 to 2 times per month. This was equally frequent in patients with a past history of either high/intermediate or low malformations.

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		High/Intermediate n (%)	Low n (%)	p	Total n (%)
Constipation (n=61)	Never/Rarely/Sometimes	23 (85.2)	30(88.2)		53(86.9)
	Often/Always	4 (14.8)	4(11.8)	1.00	8(13.1)
Infrequent stool (in past year) n (%) (n=60)	Yes	1 (3.7)	11 (34.4)	0.00	12 (20.3)
If infrequent stools, number of episodes in last 3 months (n=9)		3.0 ± (-)	2.8±0.9	0.91	2.9±0.9
Hard stools (in past year) n (%)(n=60)	Yes	15 (53.6)	19 (59.4)	0.58	34 (56.7)
If hard stools, number of episodes in last 3 months (n=26)		5.5 ± 6.5	3.5 ± 3.3	0.35	4.5 ± 5.1
Difficulty in evacuating stool (in past year)	Yes	20 (71.4)	19 (59.4)	0.33	39 (65)
n (%) (n=60)					
If difficulty in evacuating stool, number of episodes in last 3 months (n=20)		7.0 ± 7.7	5.4 ± 7.1	0.63	6.1 ± 7.2

Table 4. Constipation data for interviewed patients.

Enuresis (n=56)

36% (20/56) of patients had enuresis at least once in the last 3 months. Nine had daytime enuresis, 8 had nocturnal enuresis and 3 had enuresis both during the day and at night. 16% of patients with daytime enuresis and 13% with night-time enuresis had an episode within the last week.

Patients with either daytime or nocturnal enuresis were younger than those who did not have enuresis (daytime enuresis 7.9 ± 5.0yr vs no enuresis 12.7 ± 6.3yr, $p<0.05$; nocturnal enuresis 9.0 ± 3.7yr vs no enuresis 13.1 ± 6.6yr, $p<0.05$).

		High/Intermediate n (%)	Low n (%)	p	Total n (%)
Diarrhoea (n=57)	Never/Rarely/Sometimes	20(76.9)	30(96.8)		50(87.7)
	Often / Always	6(23.1)	1(3.2)	0.04	7(12.3)
Excoriation (n=52)	Never	17(68)	26(96.3)		43(82.7)
	Sometimes	5(20)	1(3.7)		6(11.5)
	Always	3(12)	0(-)	0.02	3(5.8)
Rectal prolapse (n=59)	Yes	1(3.7)	0(-)	0.46	1(1.7)
Abdominal pain (n=60)	Yes	21(75)	21(65.6)	0.43	42(70)
Abdominal pain (frequency) (n=32)	1-6 times in past yr	6(37.4)	7(43.8)		13(40.6)
	1-2 times in past month	5(31.3)	4(25)		9(28.1)
	1-3 times in past wk	5(31.3)	5(31.2)	1.00	10(31.3)

Table 5. Diarrhoea data for interviewed patients.

Continence Aids (n=58)

29% (17/58) of patients used continence aids for soiling (6/17), enuresis (6/17) or both (5/17). Aids were used more frequently at night (12/58) than during the day (9/58). Daytime use was primarily for soiling, however nocturnal use was primarily for enuresis.

Patients with high/intermediate malformations were more likely to use aids at night (high/intermediate 8/26 vs low 4/32, $p<0.05$). Those who used aids overnight were younger than those who did not use them (continence aid use 7.7 ±3.6yr vs no use 12.7 ± 6.7yr, $p<0.05$).

Diet (n=61)

Over half (35/61) of patients reported adverse effects after some foods. Of these, 14 (40%) reported problems with fruit, 11 (31%) with vegetables, 16 (46%) with dairy, 12 (34%) with grains/breads/cereals, 13 (37%) with fatty or fast foods, 1 (3%) with meat, and 8 (23%) with other foods.

Overall, the most commonly reported adverse effects were diarrhoea (60%), constipation (28%) and abdominal discomfort (5%). Symptoms after fruit included diarrhoea (9/14, 64%), constipation (4/14, 29%) and peri-anal discomfort (2/14, 14%). Ingestion of vegetables was associated with diarrhoea (9/11, 82%) and constipation (1/11, 9%). Symptoms after dairy included diarrhoea (6/16, 38%), constipation (8/16, 50%) and abdominal discomfort (2/16, 13%). Grains/breads/cereals induced diarrhoea (7/12, 58%), constipation (3/12, 25%) and abdominal discomfort (1/12, 8%). Symptoms after fatty or fast foods included diarrhoea (7/13, 54%) and constipation (5/13, 38%). One patient experienced diarrhoea after meat.

54% (33/61) of patients either partially or totally restricted some foods from their diet because of these adverse effects. The most frequently restricted food groups were dairy (22%), fatty or fast foods (21%), fruit (17%) and vegetables (16%).

Medication (n=63)

Over half (34/62) of patients had used a medication or other treatment for bowel related problems in the past year. 29 patients used laxatives, 2 used anti-diarrhoeals and 10 used bowel washouts. Most (27/34) patients were using some form of treatment at the time of interview. Medication-users at the time of interview were significantly younger than those who did not use medication (current medication use 9.2 ± 4.7yr vs no medication 10.4 ± 5.3yr, $p<0.05$). Of those who were using medication, 12 were using more than one type of laxative. Those with a high/intermediate malformation were more likely to be using a medication (high/intermediate 20/28 vs low 14/34, $p<0.05$). Patients with soiling were also more likely to be using medication (soiling 27/44 vs no soiling 5/16, $p<0.05$). Those who used laxatives were significantly younger than those who used anti-diarrhoeals (mean age: laxative 8.9 ± 4.3yr vs anti-diarrhoeal 19.9 ± 2.4yr, $p<0.05$).

Physical and Social Aspects

6% (4/62) of patients had limited their physical activities because of soiling or odour.

Soiling interfered with social activities of 11 (19%) patients. Of these, 9 had some parental or self-imposed restrictions and 2 had extreme limitations. As expected, patients with soiling were more likely to have social limitations than patients without soiling (social limitation: soiling 11/40 vs no soiling 0/16, $p<0.05$). Odour from soiling was also responsible for interference with social activities in 14% (8/58) of patients. Of these, 5 patients were rarely affected and 3 were frequently affected.

When questioned on the level of dependency on toilet facilities, 3.5% (2/57) of patients had used toilet facilities at least once every 30 minutes, and 5% (3/57) had to use toilet facilities at least once every hour.

On average, patients had been absent from school 0.8 ± 2.4 days in the past two school terms.

Two of the 9 patients who were in part-time or full-time work reported some limitations to their employment because of bowel dysfunction.

Health Care (n=62)

58% (34/59) patients had not had a follow-up visit for their bowel in over a year. Of these, 15 had not had a follow-up visit in the past 5 years. Patients who had follow-up visits within the last year were younger (<1yr follow-up: young 0-11yr 20/39 vs old 12-33yr 5/20, $p<0.001$) and have a high/intermediate malformation ($p<0.05$). Patients with soiling were more likely to have had a follow-up visit in the past year (<1yr follow-up: soiling 19/41 vs no soiling 4/16, $p<0.05$).

12% (6/49) of patients who attended school had an integration aid for bowel related problems (ie. cleaning after soiling).

37% (23/62) of patients received financial assistance from the Commonwealth Government because of health issues (Health Care card). Patients with high/intermediate malformations

(Health Care card: high/intermediate 15/28 vs low 8/34, $p<0.05$) and patients with soiling were more likely to have a health care card (Health Care card: soiling 21/44 vs no soiling 2/16, $p<0.05$) for bowel related problems.

27% (17/62) of patients received financial assistance in the form of a Disability allowance or their families received Carer's allowances. Patients with soiling were more likely to have received Disability allowances (Disability allowance: soiling 16/54 vs no soiling 1/16, $p<0.05$) for bowel related problems.

Continence and Quality of Life Scores

Templeton continence scores could only be determined for 23% (14/62) of patients. Based on the Templeton classification, 3 patients had "poor", 6 "fair" and 5 had "good" continence.

The requirement for digital anal examination meant that Holschneider classification of continence was able only used in 23% of patients. Four were classified as having "fair" continence, 7 "good" and 3 "normal".

Classification of quality of life using the Ditesheim and Templeton scale was valid for 85% (53/62) of patients. Five patients had "fair" quality of life and 48 "good".

Manometry (n=12) (Table 6)

Baseline anal sphincter pressures were significantly less than normal reference ranges, but there was little difference between those who did and did not soil. Recto-anal inhibitory reflexes were present in 5 patients, but the threshold volume for eliciting the reflex was significantly greater than normal. Sensation to rectal distension was also blunted. Most were unable to evacuate a water filled rectal balloon normally.

Down's Syndrome Group (n=3)

The three patients with Down's syndrome and high/intermediate anorectal malformations were 12 ± 3.4 years old. Two were male and one female. Two had a redo PSARP operation. Co-morbidities included cardiac, gastrointestinal (non colorectal), renal and respiratory.

All three had a bowel action each day. Stool consistency was formed in two and pasty in one patient. Two patients could hold back stool more than 75% of the time.

All three had soiled within the past year. Three "often" had daytime soiling episodes. Daytime soiling varied between a pasty to formed consistency and streak to large amount. One "often" soiled nocturnally and two wore nappies at night.

Only one patient had constipation and abdominal pain. The other two had episodic diarrhoea, and one also had anal excoriation.

One patient had both day and night enuresis and one only had daytime enuresis in the last week.

One patient used incontinence aids day and night for enuresis and soiling and another also used incontinence aids for enuresis and soiling but only at night.

	Soiling Never//Sometimes	Soiling Often/Always	p	Total	Historical normal values	p
Baseline sphincter pressure (mean±SD)	38.1±20.8	37.9±20.8	0.99	38±19.4	53±12	<0.01
Recto-anal inhibitory reflex						
• Normal	1	1		2		
• Present but abnormal	1	2		3		
• Not present	1	4	0.64	5		
Recto-anal inhibitory reflex (if present) – threshold (mL)	35±7.1	32.5±15	0.84	33.3±12.1	16±7	<0.01
Sensation to distension (mL)	69±79.9	90±65.4	0.62	80.5±69.4	14±7	<0.01
Co-ordination						
• Relaxation	2	1		3		
• No change	2	2		4		
• Anismus	0	2	0.33	2	10%	
Ability to evacuate water-filled balloon						
100mL	1	1		2	100%	
50mL	1	2		3		
30mL	0	0		0		
Unable	2	4		6		

Table 6. Manometry results from 12 patients.

No patient had any limitations to their physical activities but one did limit social activities because of odour from soiling, and one needed access to toilet facilities every 30 minutes.

All patients had a health care card and received disability allowance.

Only two patients had a follow-up for their bowel in the past 12 months. One was using laxatives at the time of interview for bowel control and another patient had used laxatives in the past year but had ceased 4 months prior.

All three had adverse effects from foods. These foods included fruit, vegetables and fatty or fast foods. The effects from these foods included diarrhoea and abdominal discomfort.

Social impact of incontinence – parental free comments (overall group of 62)

Earlier diagnosis:

- One parent felt as though diagnosis (of ARM) could have been made sooner if they (parents) had been more assertive. It took a day to diagnose although the Mother knew that something was not right with her son after birth, despite the nurses saying that he was fine.

Genetics:

- Two mothers thought there might be a genetic link. One mother was concerned about her future pregnancies. Two mothers were also concerned about the chance of their children's children having malformations.

Social limitations:

- One patient was teased at school because she wore a nappy and this would really upset her. Two patients were limited in their social activities because they were embarrassed to sleep over at friends or did not go to camps because of inadequate toilet facilities. School was difficult for one boy because he had to go to the toilet quite frequently during class.

Coping:

- Parents and patients coped with the disease in various ways. Three parents said that you just have to deal with it, with one adding that that was her role as a mother. One family put the disease into perspective for their child by saying that it could be a lot worse. One parent felt that being open and not making a big deal would hopefully decrease the chance of problems in the future. One mother said that you "have to have a sense of humour or you just go nuts".

- One parent said that she had to block out how bad the anal dilatations were in order for her to perform them. Two patients said that they had learnt to deal with it, with one stating that there was no need to dwell on it and another saying that you would cope with whatever happened when it came. One mother said that at times her daughter was quite upset with her malformations and procedures, however she understands their need. One patient felt that there is nothing he can do so he just deals with it. One patient always asks his parents why it has happened to him. One patient used to ask why this happened to her and feel really helpless.

- One parent said that the disease affects the whole family, not just the patient. Three parents were quite distressed about having to give their children treatment (medication, bowel washouts or anal dilatations). One parent said that it was upsetting to see their child go through it all. One mother said that it was "mentally destroying" to hear her baby crying all the time when he was passing bowel actions. She said that she did not know how she coped without any help. One patient developed a fear of going to the toilet because he would be in so much pain.

Associated disabilities:

- There were some questions by both parents and patients surrounding physical ability and if this was the reason for soiling.

Medical interactions:

- A few parents wanted more follow-up visits to occur and at more frequent times so they could monitor their child's progress. One parent did not feel that there was a need to see her doctor because he did not offer her any new information.

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4. Discussion

This review reaffirms the importance of problems with bowel control in the long term outcome after surgical repair of anorectal malformation. Over two thirds of patients had faecal incontinence over the past year and half of these had soiling which occurred either often or always. Half of all the patients soiled both during the day and at night. As expected, patients with high/intermediate malformations were more severely affected. The most severe "heavy" soiling occurred in approximately a fifth of the patients. These patients were more likely to be male or have high/intermediate malformations.

There was evidence that bowel control improved with age, but faecal urgency was still a problem in half of those aged 12 years and over. Children with low malformations tended to be less severely affected but there was also a suggestion of a slightly different symptom complex. Constipation was more common in patients with low malformation and diarrhoea more common in patients with high/intermediate malformations. Those with Down syndrome also had a poor outcome but we had no direct comparison group for this small number of children.

Only one quarter of potentially eligible patients in our study were surveyed. It is possible that this may have skewed the results toward a poorer outcome. Certainly, some studies do show rather better outcomes, with only 20% having faecal incontinence using some scoring systems [17]. However, other studies have shown broadly similar long term outcomes for continence and quality of life [18-21].

The long term functional impact in adults upon continence and quality of life can be devastating. Rintala et al. examined the outcome for adults (mean age 35 years) in terms of quality of life following surgical repair high or intermediate anomalies compared to low anomalies [22, 23]. This pair of controlled studies determined that good continence was present in only 18% of patients with high or intermediate anomalies and in 60% of those with low anomalies. Pelvic dysfunction extended into other areas. Sexual dysfunction was present in 30% of those with high or intermediate anomalies and 13% of those with low anomalies.

Further operations have been offered and there is some evidence supporting a role for posterior sagittal anorectoplasty (PSARP) with or without provision of an antegrade continent enema in selected patients with persistent and unresponsive soiling [24]. However, in our study, soiling persisted even in those patients who had received a PSARP, suggesting that it may improve function it does not usually return bowel function to normal.

Other therapies have been offered. Biofeedback has been suggested in uncontrolled studies to provide substantial improvement in continence after surgical repair of ARM [25-28]. This should be evaluated critically as biofeedback has shown similar promise in uncontrolled studies of childhood encopresis which disappeared when evaluated within the framework of fully case-controlled trials [29].

Enuresis was also a problem in many patients. Some of these children may have also had urinary system anomalies. Where they occur, it has been suggested that they are at least as

serious and complex as gastrointestinal anomalies, and contribute substantially to long term morbidity [17].

The social impacts of chronic gastrointestinal and urinary dysfunction are substantial. Approximately a quarter of the respondents in our study were on some form of financial health care assistance. Some of the comments made by parents would seem to go against the relatively optimistic outcomes on quality of life as determined by the Ditesheim and Templeton scale in our study, which suggested over three-quarters had a "good" quality of life. Quality of life measures can be extremely difficult to establish and there may be a case for looking more carefully at developing more specific and sensitive parameters [10].

An interesting observation was the impact of diet on bowel dysfunction. There is relatively little information on this area in the literature although an earlier study by the same authors in children with Hirschsprung disease identified similar findings in that group [14]. Many of the food groups which were problematic included either fibre-containing products (vegetables), lactose (dairy) or grains. There has certainly been a great deal of recent interest in the impact of rapidly fermentable, short-chain carbohydrates on functional bowel disease [30]. These have been given the acronym "FODMAPS", and found to have a substantial impact on functional abdominal pain, bloating and diarrhoea which are symptoms of irritable bowel syndrome [30]. Why this should be a particular problem in children after anorectal surgery such as Hirschsprung or anorectal malformation is not clear but may relate to the loss of a "rectal brake".

The assessment of residual function can be difficult. Several clinical-based protocols have been developed, but are poorly standardized with poor inter-scale correlation [31]. Anorectal manometry can identify a range of dysfunctional parameters which might be thought likely to contribute to incontinence [32]. In our study, a number of indices were markedly abnormal. Sphincter pressures were low. Sensory thresholds to rectal distension were extremely blunted. The rectoanal inhibitory reflex was often impaired, or only elicited with very high rectal distending volumes. Incoordinate defaecation was common, as was inability to evacuate a water filled balloon. What is interesting is that there was very little statistical difference between those who soiled heavily and those who had little soiling in terms of anorectal manometric parameters. It is likely that other pathophysiological "triggers" tip the balance in favour of heavy soiling.

Rapid colonic transit may be one such trigger. Ingestion of rapidly fermentable, short-chain carbohydrates is quite likely to lead to more rapid movement of luminal content with the potential for overcoming rectal control. There are varying abilities to tolerate fermentable loads, and these depend (amongst other factors) on rate of ingestion, concomitant nutrients, small intestinal processing, and colonic flora together with their metabolic activities. There is substantially more diversity in colonic flora than previously thought, with close interactions between environment, host genotype and the metabolic functioning gut microbiome [33]. If gut microbiome does play a role in faecal incontinence in this group, however minor, it represents an opportunity for therapeutic intervention.

Author details

Anthony G. Catto-Smith[1,3*], Misel Trajanovska[1,4] and Russell Taylor[2]

*Address all correspondence to: tony.cattosmith@rch.org.au

1 Dept of Gastroenterology and Clinical Nutrition, Royal Children's Hospital, Australia

2 Dept of Surgery, Royal Children's Hospital, Australia

3 Dept of Paediatrics, University of Melbourne, Australia

4 Murdoch Childrens Research Institute, Melbourne, Australia

References

[1] Cho S, Moore SP, Fangman T. One Hundred Three Consecutive Patients With Anorectal Malformations and Their Associated Anomalies. Arch Pediatr Adolesc Med. 2001 5/2001;155(5):587-91.

[2] Shaul DB, Harrison EA. Classification of anorectal malformations--initial approach, diagnostic tests, and colostomy. Semin Pediatr Surg. 1997 11/1997;6(4):187-95.

[3] Ong NT, Beasley SW. Long-term continence in patients with high and intermediate anorectal anomalies treated by sacroperineal (Stephens) rectoplasty. J Pediatr Surg. 1991 1/1991;26(1):44-8.

[4] Langemeijer RA, Molenaar JC. Continence after posterior sagittal anorectoplasty. J Pediatr Surg. 1991 5/1991;26(5):587-90.

[5] Templeton JM, Jr., Ditesheim JA. High imperforate anus--quantitative results of long-term fecal continence. J Pediatr Surg. 1985 12/1985;20(6):645-52.

[6] Holschneider AM, Pfrommer W, Gerresheim B. Results in the treatment of anorectal malformations with special regard to the histology of the rectal pouch. Eur J Pediatr Surg. 1994 Oct;4(5):303-9.

[7] Kelly JH. The clinical and radiological assessment of anal continence in childhood. Aust N Z J Surg. 1972 8/1972;42(1):62-3.

[8] Kiesewetter WB, Chang JH. Imperforate Anus: a five to thirty year follow-up perspective. ProgPediatrSurg. 1977 1977;10:111-20.

[9] Stephens FD, Durham-Smith E. Classification, identification, and assessment of surgical treatment of anorectal anomalies. Pediatr Surg Int. 1986 1986;1:200-5.

[10] Trajanovska M, Catto-Smith AG. Quality of life measures for fecal incontinence and their use in children. J Gastroenterol Hepatol. 2005 Jun;20(6):919-28.

[11] Stephens FD, Smith ED. Classification. In: Stephens FD, Smith ED, editors. Anorectal malformations in children. Chicago: Year Book Medical Publishers; 1971.

[12] Catto-Smith AG, Coffey CM, Nolan TM, Hutson JM. Fecal incontinence after the surgical treatment of Hirschsprung disease. J Pediatr. 1995 12/1995;127(6):954-7.

[13] Catto-Smith AG, Trajanovska M, Taylor RG. Long-term continence in patients with Hirschsprung's disease and Down syndrome. J Gastroenterol Hepatol. 2006 Apr; 21(4):748-53.

[14] Catto-Smith A, Trajanovska M, Taylor R. Long term continence after surgery for Hirschsprung's disease. J Gastroenterol Hepatol. 2007;22(12):2273-82.

[15] Catto-Smith AG, Nolan TM, Coffey CM. Clinical significance of anismus in encopresis. J Gastroenterol Hepatol. 1998 9/1998;13(9):955-60.

[16] Dewan PA, Elsworthy E, Mathew M, Poki O, Khaw SL, Roberts K, et al. Bowel imbrication in the management of anorectal anomalies. Pediatr Surg Int. 2004 Sep;20(9): 708-13.

[17] Senel E, Akbiyik F, Atayurt H, Tiryaki HT. Urological problems or fecal continence during long-term follow-up of patients with anorectal malformation. Pediatr Surg Int. 2010 Jul;26(7):683-9.

[18] Schmidt D, Jenetzky E, Zwink N, Schmiedeke E, Maerzheuser S. Postoperative complications in adults with anorectal malformation: a need for transition. German Network for Congenital Uro-REctal Malformations (CURE-Net). Pediatr Surg Int. 2012 Aug;28(8):793-5.

[19] Pruthi GK, Mohta A. Psychosocial burden and quality of life in parents of children with anorectal malformation. J Indian Assoc Pediatr Surg. 2010 Jan;15(1):15-8.

[20] Bai Y, Yuan Z, Wang W, Zhao Y, Wang H, Wang W. Quality of life for children with fecal incontinence after surgically corrected anorectal malformation. J Pediatr Surg. 2000 3/2000;35(3):462-4.

[21] Stenstrom P, Kockum CC, Bener DK, Ivarsson C, Arnbjornsson E. Adolescents with anorectal malformation: physical outcome, sexual health and quality of life. Int J Adolesc Med Health. 2013 May 1:1-11.

[22] Rintala R, Mildh L, Lindahl H. Fecal continence and quality of life in adult patients with an operated low anorectal malformation. J Pediatr Surg. 1992 7/1992;27(7):902-5.

[23] Rintala R, Mildh L, Lindahl H. Fecal continence and quality of life for adult patients with an operated high or intermediate anorectal malformation. J Pediatr Surg. 1994 6/1994;29(6):777-80.

[24] Ardelean MA, Bauer J, Schimke C, Ludwikowski B, Schimpl G. Improvement of continence with reoperation in selected patients after surgery for anorectal malformation. Dis Colon Rectum. 2009 Jan;52(1):112-8.

[25] Martins JL, Pinus J. Use of biofeedback (BFB) in the treatment of fecal incontinence after surgical correction of anorectal malformations by posterior sagittal anorectoplasty (PSARP). Sao Paulo Med J. 1997 5/1997;115(3):1427-32.

[26] Iwai N, Iwata G, Kimura O, Yanagihara J. Is a new biofeedback therapy effective for fecal incontinence in patients who have anorectal malformations? J Pediatr Surg. 1997 11/1997;32(11):1626-9.

[27] Rintala R, Lindahl H, Louhimo I. Biofeedback conditioning for fecal incontinence in anorectal malformations. Pediatr Surg Int. 1988 1988;3:418-21.

[28] Leung MW, Wong BP, Leung AK, Cho JS, Leung ET, Chao NS, et al. Electrical stimulation and biofeedback exercise of pelvic floor muscle for children with faecal incontinence after surgery for anorectal malformation. Pediatr Surg Int. 2006 Dec;22(12): 975-8.

[29] Loening-Baucke V. Biofeedback training in children with functional constipation. A critical review. Dig Dis Sci. 1996 Jan;41(1):65-71.

[30] Gibson PR, Shepherd SJ. Evidence-based dietary management of functional gastrointestinal symptoms: The FODMAP approach. J Gastroenterol Hepatol. 2010 Feb;25(2): 252-8.

[31] Ochi T, Okazaki T, Miyano G, Lane GJ, Yamataka A. A comparison of clinical protocols for assessing postoperative fecal continence in anorectal malformation. Pediatr Surg Int. 2011 Jan;28(1):1-4.

[32] Senel E, Demirbag S, Tiryaki T, Erdogan D, Cetinkursun S, Cakmak O. Postoperative anorectal manometric evaluation of patients with anorectal malformation. Pediatr Int. 2007 Apr;49(2):210-4.

[33] Spor A, Koren O, Ley R. Unravelling the effects of the environment and host genotype on the gut microbiome. Nat Rev Microbiol. 2011 Apr;9(4):279-90.

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