Assessment of functional outcome in patients of Hirschprung's disease following two stages Duhamel pull through

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Abstract

Purpose: The aim of this study was to assess the functional outcome in patients of Hirschprung's disease (HD) following two stages Duhamel pull through operation, that is levelling colostomy followed by the pull through.

Materials and Methods: The study was conducted in the Department of Pediatric Surgery of a busy tertiary care children's hospital. Patients who completed the two stage Duhamel pull through operation at least 6 months prior and aged more than three years were included in the study. The quality of life (QOL) was assessed by using a questionnaire to the parents and clinical examination of the child. Continence status was assessed by applying the Krickenbeck Score.

Results: Thirty patients were assessed after completion of two stage Duhamel pull through operation. Quality of life (QOL) was good in 27 patients (90%), fair in 2 patients (6.6%) and poor in 1 patient (3.3%). Voluntary Bowel Movement was present in 25 patients (83.3%) and absent in 5 patients (16.6%). Soiling was absent in 27 patients (90%) and present in 3 patients (10%). Among these 3 patients, 2 patients had grade 1 and 1 patient had grade 2 soiling. Constipation was absent in 26 patients (86.6%) and present in 4 patients (13.3%). Among these 4 patients, 1 patient had grade1 and 3 patients had grade 2 constipation.

Conclusion: In patients of Hirschprung's disease, fecal continence and the quality of life was satisfactory after the two stage Duhamel pull through operation.

Keywords: Hirschprung disease, Duhamel pull through, Continence, Quality of life.

Introduction

Hirschsprung's disease is a well known disease of childhood. Several studies on the frequency of HD have been reported. The incidence of HD is estimated to be 1 in 5000 live births and ranges from 1 in 4,4,00 to 1 in 7,000 live births.[1,2] It has long been recognized that males are more commonly affected than females with a male: female ratio of 4:1[3,4] The male preponderance is less evident in long segment HD where the male: female ratio is 1:1-2:1 and is even reversed in total colonic aganglionosis, where the male: female ratio is 0.8:1.[1] Therapeutic options for Hirschsprung's disease have gradually become refined through trial and error. The first treatment consisted of a diverting colostomy, which relieved the child's symptoms; however, symptoms returned after closure of the ostomy.[5] Attempts at bypass or removal of the redundant portions of the colon were uniformly unsuccessful. One of the more intriguing approaches was a lumbar sympathectomy, or spinal anaesthesia.[6,7] Theoretically removal of the sympathetic input to the distal rectum would result in predominance of parasympathetic or relaxation impulses. Several patient had an improvement in symptoms with this approach, but the procedures was eventually condemned by Ladd. Of the surgical treatment of HD Swenson and Bill are credited with the first successful surgical approach.[8] A number of operative procedures have been described in relation to HD ranging from single stage transanalendorectal pull through to staged procedures which basically depends on the surgeon's preference and the condition of the patient. Each procedure however has its own advantages and disadvantage. Duhamel first described his operation for H.D. in 1956. The procedure consisted of a retrorectal dissection, whereby a significant portion of the aganglionic rectum was preserved and anastomosed to ganglionated proximal colon.[9,10] The advantage of this procedure includes ease of performance, reduction of anastomotic leak and strictures, retention of anal sensory receptors and preservation of nervi-erigentes. Duhamel devised his operation as a simplified variation of Swenson rectosigmoidectomy and intended to apply it earlier in infancy. Modifications of Duhamel operation have been numerous, including placement of the anastomosis above the internal sphincter to avoid incontinence. Additional modification has involved the creation of the anastomosis with the use of mechanical stapling devices.[11] An additional modification has centred around eliminating the proximal rectal pouch "SPUR" in the aganglionic rectum, which can trap large accumulation of stool. Martin and Altemeir and Martin and Caudill described careful clamp placement to entirely eliminate the rectal pouch.[12] Use of stapling devices for the colorectal anastomosis facilitate division of the common rectal wall.

The primary pathology is the lack of ganglion cells in the myentric and sub mucosal plexus leading to a lack of propagation of the peristaltic wave. The cause of the spasm in the aganglionic bowel is unclear. Cholinergic hyperenervation, inadequate distribution of nitric oxide synthetase, and abnormalities of the

interstitial cells of Cajal have all been reported to be associated with Hirschsprung's disease.

Depending upon the level at which arrest takes place in foetal life, Hirschsprung's comprises of following types-

1. Short segment (classical form)-involves rectum and recto sigmoid junction.
2. Long segment HD-involves descending colon
3. Subtotal HD-involves mid transverse colon
4. Total colonic agangliosis-involves entire length of large bowel
5. Total intestinal agangliosis-involves up to ileum and even up to jejunum.

In our institute most common practicing surgery for Hirschsprung's disease is two stage Duhamel pull through.

Assessment of Long Term Outcomes: As done in most diseases the long term outcomes of the operated cases of Hirschsprungs disease could be assessed under following heads.

1. Clinical assessment.
2. Clinical continence scoring system.
3. Quality of life

Clinical Assessment: Includes the assessment of the general physical growth of the patients presence or absence of stunting. Postoperative clinical assessment also includes examination of the wound, assessment for mucosal prolapsed, gluteal symmetry, presence of obvious fistula or sinuses in the perineum, strictures, constipation and incontinence. Digital examination is very important in cases of Hirschsprung disease, with a proper digital examination one can assess and predict the level of continence, eg; a good squeeze pressure on command is associated with a good result besides this, presence of fecaloma, loaded rectum indicated the presence of constipation or incomplete emptying, presence of stricture and posterior rectal shelf can also be appreciated with a good per rectal examination

Clinical Continence Scoring Systems: Most of the continence scoring systems were originally described in relation to the anorectal malformations but they still hold rule for Hirschsprung disease. Standardized assessment of clinical outcome after repair of HD is essential for appropriate quality control in series of patents treated in single or different institution, and for comparing different treatment modalities. Clinical assessment is subjective and may be biased by the observer, who is often the surgeon treating the patient. Various scoring systems have been in vogue.[7] For any scoring system to be successful it needs to be simple, easy to use comparable and should be able to differentiate between subtle grades of continence. None of the scores have undergone a proper validation process explaining the fact why none of these scores are universally accepted.

There is consensus that faecal continence represents the most important endpoint in patients with HD. Therefore, specific scores for assessment of long-term results are focused on differentiating various degrees of faecal incontinence. No consensus has been achieved on including and scoring other symptoms such as constipation, urinary incontinence or quality-of-life measurements.

Soiling and constipation are the two most important parameter in assessing the efficacy of the procedure and determining the long term outcome.Various scores have been used to quantify the stooling in the postoperative period. These are Kelly, Rintala, Holschneider and Pena score. The Kelly score has been widely used and has gained popularity. Each score tries to assess maximum period of time the child remains dry without soiling or incontinence. The latest is Krickenbeck score (2005) which we had followed in this study.

Quality of life (QOL) has become established as an important end point in medical care. QOL depends on numerous factors like physical and mental health, psychosocial adjustment, family and peer group support, socioeconomic status and educational achievement. Quality of life score (QOL) in the patients of Hirschsprung's disease looks into the social disability aspect in children with defective stooling and its impact on their daily activity. There is no universally accepted score. QOL score will be obtained by asking the parents to fill in the questionnaire.

Materials and Methods

The cross sectional study was conducted in department of Paediatric Surgery at Lady Hardinge Medical college and associated Kalawati Saran Children's Hospital. 30 patients was admitted, operated and followed up. Patients with age more than three year and who underwent Duhamel operation at least 6 month prior was included in the study.

Exclusion Criteria

1. Parents who refused to for participation in study.
2. Patients who had undergone one or three stage Duhamel pull through.
3. Patients who had undergone Soaves and Swenson's operation.
4. Patients who had undergone redo Duhamel pull through procedure

Outcome Measures

1. Primary measure-To assess continence status following two stage Duhamel pull through surgery.
2. Secondary measure- To assess postoperative Quality of life six months after two stage Duhamel pull through surgery.

Ethical committee clearance was taken for the study. Contact details of all patients of Hirschprung's disease operated in the department of pediatric surgery, Lady Hardinge Medical College and Kalawati Saran Children's Hospital, New Delhi fulfilling the inclusion criteria visiting outpatient department and from the

medical record department was extracted. These patients were contacted over phone and through letters and was requested to take part in the study. Parents who had refused for consent was excluded. After proper written informed consent the study was started.

Clinical Assessment: All patients were examined clinically with special attention to the nutritional status, abdominal scar, perineal wound and evidence of soiling and incontinence.

Stooling Score

Krickenbeck Score: At the Krickenbeck Meeting in 2005, consensus was achieved concerning the assessment of outcome after HD repair. The method comprises three parameters: voluntary bowel movements (Yes/ No), Soiling (Yes/ No, if yes grade 1-3), and constipation (Yes/No, if yes grade 1-3). The assessment was performed in children more than 3 years of age who were not underwent any form of therapy.

Krickenbeck score (2005)

1. **Voluntary Bowel Movements** Yes/ No

a. Feeding of urge
b. Capacity to verbalize
c. Hold the bowel movement

2. Soiling

a. Grade 1 Occasionally (Once or twice per week)
b. Grade 2 Every day, no social problem
c. Grade 3 Constant, social problem

3. Constipation

a. Grade 1 Manageable by changes in diet
b. Grade 2 Requires laxatives
c. Grade 3 Resistant to diet and laxatives

Quality of life score (QOL): The QOL score was obtained by asking the parents to fill in the questionnaire as given below. The total score was calculated at the end of the questionnaire.

QOL Questionnaire

1. Social habit

Regular habit-3
Irregular habit-2
Different habit with problem-1
No social habit-0

2. School attendance-

Full time-2
Part time-1
Never -0

3. Daily activity-

Normal as peer-2
Less than peer-1
Restricted -0

4. Relation to peer –

Good-2
Limited -1
No relation -0

5. Feeling-

Normal-3
Feeling different-2
Less appreciated-1
Afraid and depressed-0

The QOL would then be termed as
Good (8-12)
Fair (5-7)
Poor (0-4)
based on the scores achieved.

Results

A total of 30 patients were included in the study out of which 24 were males and 6 were females (M:F = 4:1). Among these 30 patients, 27 patients had rectosigmoid type of HD and 3 patients had long segment HD. Among the 3 patients with long segment HD, two patients had aganglionosis till mid transverse colon and one patient had aganglionosis till the splenic flexure.

Voluntary Bowel Movement (VBM): VBM was noted as being present only if the patient fulfilled all the three criteria i.e. feeling of urge, capacity to verbalize and ability to hold stools. VBM was present in 25 (83.33%) patients (20 males and 5 females) and absent in 5 (16.66%) patients (1 female and 4 males). Therefore 20 males (83.33%) and 5 females (83.33%) had VBM. The difference between male and female was not statistically significant (p-value-1.000).

Comparing the type of HD in relation to the VBM of these 30 patients, 27 (90%) patients had rectosigmoid type of HD and 3 (10%) patients had long segment type of HD. Among rectosigmoid type HD, 23(85.1%) had VBM present and in 4 (14.8%) patients VBM were absent. Among long segment type of HD, 2 (66.6%) had presence of VBM and 1 (33.3%) patient had absence of VBM. The difference in the two groups based on the type of HD was not statistically significant (p-value-0.433)

Soiling: There was no soiling in 27 (90%) patients while some degree of soiling was present in 3 (10%) patients. Sub classifying soiling, soiling present once/twice a week(S1) was seen in 2 (6.67%) patients, everyday not causing social problem (S2) was seen in 1 (3.33%) patients and constant causing social problem (S3) was not seen in any patient.

Soiling was present in 3 males (12.5%) and absent in 21 males (87.5%) and soiling was absent in all the females, the difference between the groups being statistically insignificant (p-value-1.000).Among patients with rectosigmoid type of HD, only 3(11.1%) patients had soiling present and of these 2 had S1 grade of soiling and 1 had S2 grade of soiling. Soiling was absent in patients with long segment HD.

Constipation: Constipation was present in only 4(13.33%) patients.constipation requiring dietary modification (C1) was seen in 1(3.33%) patient, constipation requiring laxatives (C2) seen in 3(10%) patients and constipation resistant to both diet and laxative (C3) was not present in any patient.

Constipation was present in 3 males (12.5%) and absent in 21 males (87.5%); constipation was present in 1 female (16.66%) and absent in 5 females (83.33%). Among the 27 patients with rectosigmoid type of HD, 3(11.1%) patients had constipation present and 24 (88.88%) patients had constipation absent. Among long segment HD, one patient (33.3%) had constipation present and 2 patients (66.6%) had constipation absent.

Table 1: Shows the incidence of the three parameters assessedfor continence i.e. presence/absence of VBM, Soiling, Constipation

		n	%
Voluntary bowel movement		25	83.33%
Soiling	Grade 1	2	6.67%
	Grade 2	1	3.33%
Constipation	Grade 1	1	3.33%
	Grade 2	3	10.00%

Assessment of quality of life (QOL) score

QOL scoring was done with a 5 point questionnaire, namely social habits, school attendance, daily activity, relation to peers and feelings, with a maximum of 12 and a minimum of 0. QOL score of 0-4 was termed as poor, 5-7 as fair and 8-12 as good.

Table 2: QOL distribution (n=30)

QOL	n	%
Poor	1	3.33%
Fair	2	6.67%
Good	27	90.00%
Total	**30**	**100%**

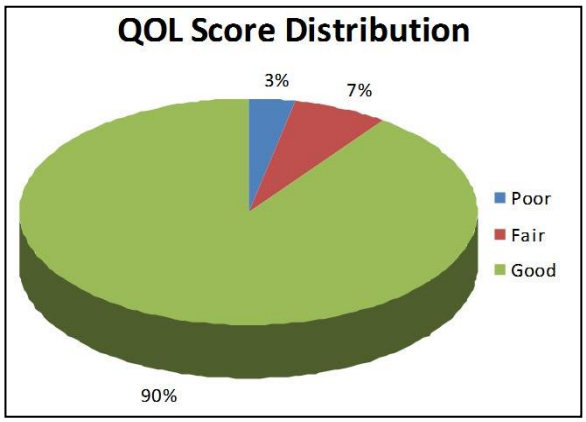

Fig. 1: QOL distribution (n=30)

Among 30 patients, 27(90%) patients had good QOL, 2 (6.67%) patients had fair QOL and 1(3.33%) had poor QOL. In terms of social habit, there were 22 patients who had regular habit, 7 patients who had irregular habit and only 1 patient had different habit with problem. 6 patients had never attended school, 17 were attending part time school and 7 attended school regularly. Daily activity was less than peers in 18 patients and remaining 12 patients had normal daily activity. Relation to peers was limited in 1 patient and good in remaining 29 patients. Feeling was normal in 24 patients, 3 patients had different feeling and rest 3 patients had unappreciated feeling. Among patients with rectosigmoid HD, 24(88.8%) patients had good QOL, 2 (7.4%) patients had fair QOL and 1 (3.7%) patient had poor QOL. All patients with long segment HD had good QOL.

Number of patients and Assessment Period from time of Definitive Surgery: Among 30 patients, 7 (23.3%) patients were examined after 6 months to 1 year of definitive surgery (group-A), 14 (46.6%) patients after 1-3 year of surgery (group-B), 7(23.3%) patients after 3-6 years of surgery (group-C) and only 2(6.67%) patients after 6-9 years of surgery (group-D).

Table 3: Number of patients and assessment period from time of definitive surgery

Time period(years)	n(%)
6months-1 (group-A)	7(23.3%)
1-3(group-B)	14(46.6%)
3-6(group-C)	7(23.3%)
6-9(group-D)	2(6.67%)

Relationship between VBM and Assessment Period from time of Definitive Surgery: Among group A,5 (71%) patients had VBM present and 2(29%) patients had VBM absent. Among group B,13 (92%) patients had VBM present and 1(8%) patient had VBM absent. Among group C,5 (71%) patients had VBM present and 2(29%) patients had VBM absent. Among group D, all (100%) patients had VBM present.

Table 4: Assessment period from time of definitive surgery and VBM

Time period (year)	n %	VBM+	VBM-
6 month-1	7(23.3%)	5(71%)	2(29%)
1-3	14(46.6%)	13(92%)	1(8%)
3-6	7(23.3%)	5(71%)	2(29%)
6-9	2(6.67%)	2(100%)	0

Chi-square p-value = 0.433

Relationship between soiling and assessment period from time of definitive surgery.

Among group A, 5(71%) patients had soiling absent and 2(29%) patients had soiling present. Among group B,13 (92%) patients had soiling absent and 1(8%) patient had soiling present. Among group C and group D, all (100%) patients had soiling absent.

Table 5: Assessment period from time of definitive surgery and soiling

Time period (year)	n%	Soiling-	Soiling+
6 month-1	7	5(71%)	2(29%)
1-3	14	13(92%)	1(8%)
3-6	7	7(100%)	0
6-9	2	2(100%)	0

Chi-square p-value = 0.283

Relationship between constipation and assessment period from time of definitive surgery.

Among group A, all (100%) patients had constipation absent. Among group B, 12 (85%) patients had constipation absent and 2(15%) patients had constipation present. Among group C,5 (71%) patients had constipation absent and 2(29%) patients had constipation present. Among group D, all (100%) patients had constipation absent.

Table 6: Assessment period from time of definitive surgery and Constipation

Time period (year)	n	Constipation- n (%)	Constipation+
6 month-1	7	7(100%)	0
1-3	14	12(85%)	2(15%)
3-6	7	5(71%)	2(29%)
6-9	2	2(100%)	0

Chi-square p-value = 0.423

Relationship between QOL score and assessment period from time of definitive surgery.

Among group A, all (100%) patients had good QOL score. Among group B,12 (83%) patients had good QOL score, 1 (8.3%) patient had fair QOL score and 1(8.3%) patient had poor QOL score. Among group C,6 (85.6%) patients had good QOL score and (14.2%) patient had fair QOL score. Among group D, all (100%) patients had good QOL score.

Table 7: QOL score and assessment period from time of definitive surgery

Time period (year)	n	QOL Good n (%)	QOL Fair	QOL Poor
6 month-1	7	7(100%)	0	0
1-3	14	12(83%)	1(8.3%)	1(8.3%)
3-6	7	6(85.6%)	1(14.2%)	0
6	2	2(100%)	0	0

Chi-square p-value = 0.868

Relationship between VBM and QOL score.

Among 30 patients, 100% patients with poor QOL score had VBM absent.50% patients with fair QOL score had VBM absent and 50% had VBM present. 88.8% patients with good QOL score had VBM present and 11.1% patients with good QOL score had VBM absent.

Table 8: QOL score in relation to VBM

Q.O.L Score	voluntary bowel movement		Total
	No	Yes	
Poor	1(100%)	0	1
Fair	1(50%)	1(50%)	2
Good	3(11.1%)	24(88.8%)	27
Total	5	25	30

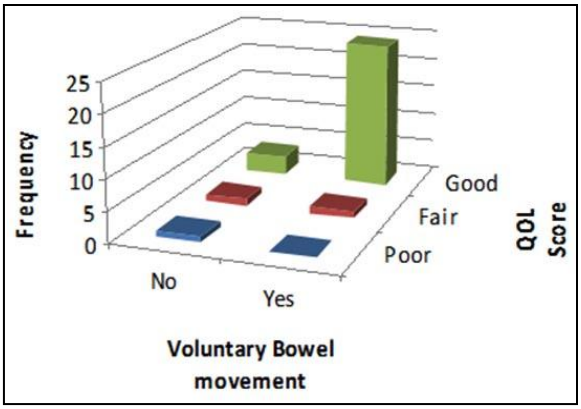

Fig. 2: QOL score in relation VBM

Chi square value = 0.027

Relationship between constipation and QOL score;

Among 30 patients, 100% patients with poor QOL score had constipation present, 50% patients with fair QOL score had constipation present and 50% had constipation present. 92.5% patients with good QOL score had constipation absent and 7.4% patients had constipation present.

Table 9: QOL score in relation to constipation

Q.O.L Score	Constipation		Total
	No	Yes	
Poor	0	1(100%)	1
Fair	1(50%)	1(50%)	2
Good	25(92.5%)	2(7.4%)	27
Total	**26**	**4**	**30**
Chi-square p-value = 0.008			

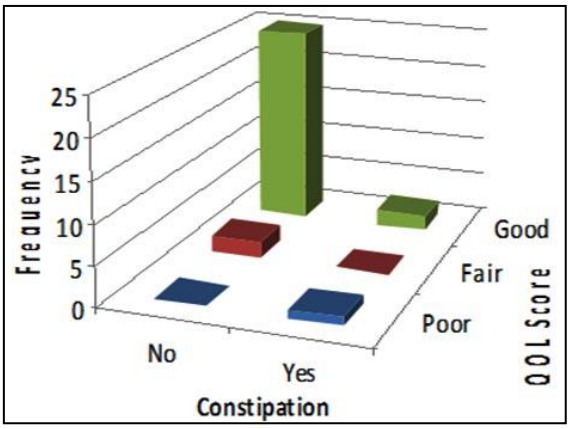

Fig. 3: QOL score in relation to constipation

Relationship between soiling and QOL score;
Among 30 patients, 100% patients with poor QOL score had soiling present, 100% with fair QOL Score had soiling absent. 92% patients with good QOL score had soiling absent and 8% patients with good QOL score had soiling present.

Table 10: QOL score in relation to soiling

Q.O.L Score	Soiling		Total
	No	Yes	
Poor	0	1(100%)	1
Fair	2(100%)	0	2
Good	25(92%)	2(8%)	27
Total	**27**	**3**	**30**

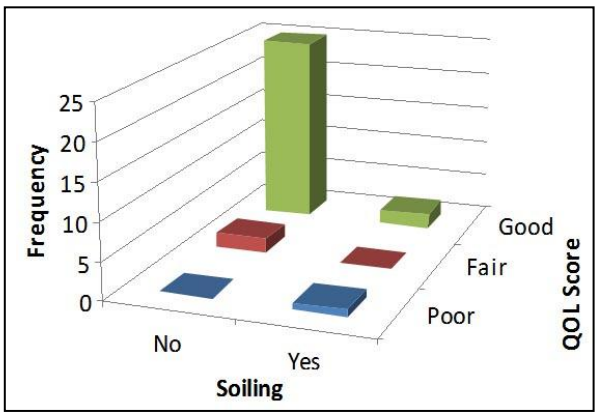

Fig. 4: QOL score in relation to soiling

Discussion

Over the years, various pull through surgeries have been used to treat patients with HD.[1-6] It has been shown in several studies that there is no statistically significant difference in the functional outcome with respect to bowel function between the various pull through surgeries.[7-11] As with many surgery, for HD also has some long and short term complications. Faecal incontinence and constipation remains major postoperative complication that impede social and psychological developmental in these patients. There is a great variation in the literature regarding functional results. The main problem in comparing different series is the highly variable criteria used in the evaluation. Most reported series have utilized some form of questionnaire for evaluation.The evaluation of quality of life using such questionnaire which are answered by the parents, may not always give the correct picture. Therefore, the final outcome may not be fully assessed until the patient have reached an age to evaluate the social consequences of possibly defective bowel control. A number of clinical scoring system are currently in use to assess the functional and stooling pattern of children with ARM and HD.[12] The stooling outcome that we employed in this study were done using the recently proposed Krickenbeck scoring system. Though this was proposed for anorectal malformation, it has also been used in the evaluation of cases of HD.[13]

In our institute, the most common practicing surgery for HD is two stage Duhamel pull through. In this study, 90% patients had recto sigmoid HD and 10% patients had long segment HD.

Voluntary Bowel Movement: VBM were considered to be present when a child had capacity to verbalize, urge to defecate and could hold his bowel movement. Jarvi et al reported that 40% patients had the ability to hold back defecation 20% had the feeling of urge to defecate.[14] Rintala et al (2012) reported that incidence of faecal soiling more frequently than once a week was13%, faecal accidents more often than once a week was 2.2% and constipation treated with laxatives or enema was 10%.[15]

In the present study, VBM was present in 83.3% patients and absent in 16.6% patients. The difference in incidence of VBM present or absent with respect to sex (p-value-1.000), type of pull through (p-value-0.433), age at evaluation (p-value-0.418) and assessment period from time of definitive surgery (p-value-0.433) was done and was not significant. This was similar to the findings of other followup studies on patients of HD managed by two stage Duhamel procedure.[16,17]

Soiling: Faecal incontinence is psychosocially the most catastrophic sequel of any type of surgery for HD. The main physiological prerequisites for faecal incontinence include preserved sphincter function, rectal reservoir, and anal canal sensation. The occurrence of different

degrees of faecal incontinence in patients who had undergone repair of HD is not unexpected.Patients with HD lack rectoanal relaxation reflex not only after but also before surgery, consistent with primary defects in the innervation and function of the internal anal sphincter.[18,19] Soiling and incontinence has been interchangeably used in various publications.While in some any soiling is a component of incontinence, in others, patient is called incontinent if there is soiling all the time.

Soiling in the initial postoperative months is a common problem as most patients following resection tend to have large volume of liquid intestinal contents. Passage of flatus in this situations invariably causes some soiling. Gradually the intestinal contents become more solid and soiling is eventually eliminated. The reported incidence of fecal incontinence is 2-20% for Swenson procedure and 1-14% for modified Duhamel procedure.[16,20-23] Diseth et al reported that incidence of soiling following two stage Duhamel pull through is 36%.[24]

In this study soiling was present in 3/30 (10%) and these results are similar to the results reported in other series which shows the incidence of postoperative soiling to range from 1-14%.[25,26] However Minford et al reported a higher incidence of postoperative soiling of 51% using the An analogue scoring system (modified Shankar et al score).[27] Among these 10% patients, 6.67% patients had grade 1 (occasionally, once or twice per week) soiling and 3.36% patients had grade 2 constipation (every day but no social problem).None had grade 3 soiling (constant, with social problem). The difference in the incidence of soiling with respect to sex(p-value-1.000), age at evaluation(p-value-0.750), type of pull through (p-value-1.000) and assessment period from time of definitive surgery (0.283) and was insignificant.

Constipation: Abnormalities in the innervation and function of the pulled through colon and internal anal sphincter may contribute to development of constipation in HD.[28] Most series reported that constipation in patients with HD become milder or subsides with the time after early childhood. Rintala et al reported that after Duhamel procedure incidence of constipation is 30% and 10% treated constipation with laxatives or enema.[15] The incidence of constipation after HD surgeries has been reported as 6-15% after Swenson procedure and 4-10% after Duhamel surgery.[16,20-23]

There are three main factors in the causation of postoperative constipation

1. Until the sensation of faeces in the neorectum is appreciated by the child he/she will ignore it and therefore, fail to empty the terminal bowel.To avoid this the patients must undergo intensive toilet training in the initial few postoperative weeks.
2. The colon proximal to the point of resection may be considerably dilated and likely to empty incompletely due to colonic inertia.
3. Distal resection should include partial sphincterotomy to take care of internal anal achalasia.

The incidence of constipation in this study was 4/30(13.3%) comparable to literature Diseth et al reported the incidence of constipation to be 16%.[29] Constipation requiring dietary modification (C1) was seen in 1 (3.33%) patient, constipation requiring laxatives (C2) seen in 3(10%) patients and constipation resistant to both diet and laxative (C3) was not present in any patients. The incidence of constipation with respect to sex (p-value-1.000), age at evaluation (p-value-0.354), type of HD (p-value-0.277) and assessment period from the definitive surgery (p-value-0.423) was done and was not significant.

Quality of Life Score: As paediatric surgeon we have great responsibilities not only in regard to surgical procedure but also in following up with these patients. Quality of life (QOL) has become an important issue in the medical community. It is important in the reporting of long term outcomes of treatment of any condition. Many patients who undergo Duhamel pull through complain of bowel function disorder. This may have a significant influence on their QOL.

To assess QOL in children is a challenge as the child's vocabulary, language, and perception of health and illness are continuously evolving. Various QOL questionnaires are available which are taken from the parents. It looks as good from the parent's perspective but may not be good for the patients.

In this study QOL scoring was done with a 5 point questionnaire, namely social habit, school attendance, daily activity, relation to peers and feelings, with a maximum of 12 and a minimum of 0.QOL score of 0-4 was termed as poor, 5-7 as fair and 8-12 as good.

Among 30 patients, 7(90%) patients had good QOL, 2(6.67%) patients had fair QOL and 1(3.33%) had poor QOL. The incidence of QOL with respect to age at evaluation (p-value-0.976), sex (p-value-0.659), type of HD (p-value-0.831) and assessment period from time of definitive surgery (p-value-0.868). These results are similar to the result of the study reported by Hartman et al who assessed QOL in postoperative patients of HD by TAC QOL questionnaires (TNO-AZL Child quality of life questionnaires) and found that the relationship between disease specific functioning and QOL remains unclear.[30] However the study reported by Gunnarsdottir et al found that QOL score were lower in females and that longer the aganglionic segment, the greater its adverse impact on QOL in later life.[31] In terms of social habit,in our study there were 22 patients who had regular habit, 7 patients who had irregular habit and only 1 patient had different habit with problem. After analysing school attendance we

found that6 patients had never attended school, 17 were attending part time school and 7 attended school regularly. The relation of school attendance with age at evaluation (p-value 0.232), constipation (p-value 0.203) and soiling (p-value 0.094) was assessed and was not found to be statistically significant. The only significant correlation was found between school attendance and VBM (p-value 0.039) which was nearly similar to the result of the study reported by Bai et al which found that school absence occurred in 13% patients because of poor continence.[32] Daily activity was less than peers in 18 patients and remaining 12 patients had normal daily activity. Relation to peers was limited in 1 (3.33%) patient and good in remaining 29 (96.6%) patients. In our study there was no significant correlation between age at evaluation (p-value 0.829), soiling (p-value 0.100), constipation (p-value 0.133) and VBM (p-value 0.167). These results are dissimilar to those from the study reported by Bai et al which found that the incidence of peer rejection was high at 15.6%.[32] Feeling was normal in 24 (80%) patients, 3 (10%) patients had different feeling and rest 3 (10%) patients had unappreciated feeling. We found that there was significant correlation between feeling and both constipation (p-value 0.015) and VBM (p-value 0.027)

After analysing the relationship between overall QOL and VBM, we found that all 1(100%) patients with poor QOL score had absent VBM. 1 (50%) patient with fair QOL score had VBM absent, and 1 (50%) had VBM present. 24 (88.8%) patient with good QOL score had VBM present while 3 (11.1%) patient with good QOL score had VBM absent. There was significant correlation between VBM and QOL (p-value 0.027).

Similarly relationship between QOL and soiling, we found that 1 (100%) patient with poor QOL had soiling present. 2 (100%) patient with fair QOL had soiling absent, 25 (92%) patient with good QOL had soiling absent and 2 (8%) patient with good QOL had soiling present. There was significant correlation between overall QOL and soiling (p-value 0.009).

After analysing relationship between constipation and overall QOL we found that 1(100%) patient with poor QOL score had constipation present, 1 (50%) patient with fair QOL score had constipation present, 25 (92.5%) patient with good QOL score had constipation absent and 2 (7.4%) patient had constipation present. There was a significant co relation between QOL and constipation (p-value 0.008).

The above finding of statistically significant correlations (VBM Vs overall QOL, soiling Vs QOL, and constipation Vs QOL) are similar to the results of the study reported by LEIRI et al and Bai et al.[32,33] Both these studies found that bowel dysfunction impairs QOL. However, Hartman et al reported that he relationship between disease specific functioning and the overall QOL in postoperative patients of HD remains unclear.[30]

In conclusion patient with HD managed by the two stage procedure of preliminary levelling end-colostomy with Hartmans procedure in the first stage, followed by the Duhamel procedure in the second stage, have good results on assessment of postoperative continence levels as well as overall quality of life. These results have no significant co relation with the gender, the age at assessment, the type of HD and the time duration from last definitive surgery. Poor continence scores (significant soiling, constipation and lack of VBM) correlate statistically with overall poor QOL especially in term of poor social habits, poor school attendance and feelings. Another significant finding is that although it is generally perceived that some postoperative problems of bowel functions improve with time and increasing age of the patients, we have not found any significant difference. A possible explanation for this finding and lacuna of this study is a relatively small number of patient in the series. Similarly the length of aganglionosis did not significantly affect the outcome but again this finding has to be interpreted with caution as only three patients (10% of total) had long segment HD and none had TCA.

Conclusion

Patients with HD managed by two stage procedure of preliminary levelling end colostomy with Hartmans procedure in the first stage followed by the Duhamel procedure in the second stage have good results on assessment of postoperative continence levels as well as overall quality of life (QOL). These results have no significant correlation with the gender, the age at assessment, the type of HD and the time duration from last definitive surgery. Poor continence scores (significant soiling, constipation and lack of VBM) correlate statistically with overall poor QOL especially in terms of poor social habits, poor school attendance and feelings. Although not statistically significant, there is some correlation between poor continence level and poor QOL in form of daily activity and relation to peers.

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