ORIGINAL PROF-1585

HIRSCHSPRUNG'S DISEASE;

MODIFIED DUHAMEL (MARTIN MODIFICATION), A PROCEDURE OF CHOICE (A STUDY AT SHEIKH ZAYED HOSPITAL RAHIM YAR KHAN)

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ABSTRACT... Introduction: Hirschsprung's Disease is one of the most common congenital anomalies that Pediatric Surgeons manage. In spite of the various modifications of pull through procedures available, the long term functional results are less than ideal. However, Modified Duhamel Procedure is one which has relatively good functional results and that is the reason we have selected this procedure for Hirschsprung's disease in Sheikh Zayed Hospital, Rahim Yar Khan. **Objectives:** To evaluate the outcome of patients after Modified Duhamel Procedure for Hirschsprung's Disease. **Design:** This is case series study. **Settings:** Department of Pediatric Surgery Sheikh Zayed Medical College/ Hospital Rahim Yar Khan. **Period:** Seventeen cases were studied over a period of two year i.e. from November 2006 to December, 2008. **Material and Method:** Seventeen (17) cases from both sexes were operated for Hirschsprung's Disease over a period of one year and Modified Duhamel Procedure was adopted for all these cases. All patients were diagnosed cases of rectosigmoidal aganglionosis and follow up was done over a period of one year according to a comprehensive Proforma. Four parameters like normal stool evacuation, abdominal distension, soiling and stool incontinence were followed and then the results were compared with other national and international studies. **Results:** Fever 19.4% (n=04), wound infection 19.4% (n=04), vomiting 9.52% (n=02), abdominal distension 4.76% (n=01), and bleeding per rectum 4.76% (n=01), were the immediate post operative complications. Fourteen patients (82.35%), used to pass stool once daily. Abdominal distension was observed occasionally in six patients (28.57%). Soiling was seen in five patients (29.41%). Out of Seventeen, stool incontinence was seen in only three patients (17.64%). **Conclusion:** Modified Duhamel Procedure with the help of linear cutter stapler device is quite safe, easy and less time consuming. Infact Modified Duhamel is a procedure of choice for Hirschsprung's Disease.

Key words: Hirschsprung's Disease, Modified Duhamel Procedure, Constipation, Soiling, Mechanical Stapling Device.

INTRODUCTION

Hirschsprung's Disease, also called "Congenital Aganglionic Megacolon" is one of the commonest causes of neonatal intestinal obstruction.

The incidence of Hirschsprung's Disease ranges from 1 in

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in 4400 to 1 in 7000 live births[1]. The male to female ratio in patients with classic Hirschsprung's Disease is generally reported as 4:1 in favor of males[2]. Bander et al[3] calculated the risk for transmition of Hirschsprung's Disease to the relatives, in his study, brothers of patients with short segment Hirschsprung's Disease have a higher risk 4% than the sisters 1%.

Hirschsprung's Disease is characterized by an absence of ganglion cells in the nerve plexus of rectum and colon associated with dilatation of the normal proximal colon due to neurogenic obstruction. The first description of a case of congenital megacolon is credited to F. Ruysch[4], a Duch anatomist who in 1691 described a 5 year old girl who died of an intestinal obstruction.

The classic description of this condition was reported by Harold Hirschsprung[5] in 1886. He was the Senior Pediatrian at the Queen Louise Children Hospital in Copenhagen. He described two children; both had the classic clinical and anatomical characteristics of the disease. Hirschsprung's in 1904 presented another case report of 10 children and disease was described as "Congenital dilatation of colon". An understanding of Hirschsprung's Disease took several more years when different theories regarding intestinal obstruction were put forward. An appreciation that distal colon was the actual abnormality was initially advanced by Tittle[6] in 1901, who identified an absence of ganglion cells in the distal colon of a child with Hirschsprung's Disease.

In 1946 Ehrenpresis[7] was the first to appreciate that the colon became secondarily dilated because of distal obstruction.

In 1948, Whitehouse, Kernohan, Zulzer and Wilson[8], definitely documented the absence of ganglion cells of the myenteric plexus in patients with Hirschsprung's Disease. Aganglionosis typically extends to the recto sigmoid region in approximately 80% cases[9]. In about 10% cases proximal colon is involved and in remaining 10% cases entire colon with variable extension into small bowel may occur. Due to the absence of ganglion cells, cholinergic activity is increased and nonadrenergic inhibitory system is decreased that leads to contracted spastic state of the aganglionic bowel[10].

The evolution of the surgical treatment for Hirschsprung's Disease over the past 50 years has been a wide variety of techniques ranging from Swenson procedure in 1948[11] to perineal one stage pull through described by Langen et al in 1999[12]. Bernad Duhamel[13] was the first to describe his operation for Hirschsprung's Disease in 1956. The operative principal of his technique included minimal pelvic dissection, a retro rectal approach for the pull through of intestine to the anal opening, a wide anastomosis between ganglionated colon and anterioly placed rectum and preservation of anterior wall of the rectum with its nerve supply[14].

There have been numerous modifications of the Duhamel Procedure. Elimination of the common wall of the rectal pouch "spur" was the main stay of different modifications. Martin and Altemier[15] described careful clamp placement to entirely eliminate common wall of rectal pouch "spur". With the application of mechanical stapling device to the colorectal anastomosis, the division of the common rectal wall "spur" was further facilitated as reported by Ikeda[16].

Modified Duhamel procedure with the help of linear cutter stapler device was performed for Hirschsprung's disease in Pediatric Surgery Department, Sheikh Zayed Hospital Rahim Yar Khan. The aim of this study was to determine the morbidity, mortality and functional outcome of this procedure and compare it with other studies.

INCLUSION CRITERIA

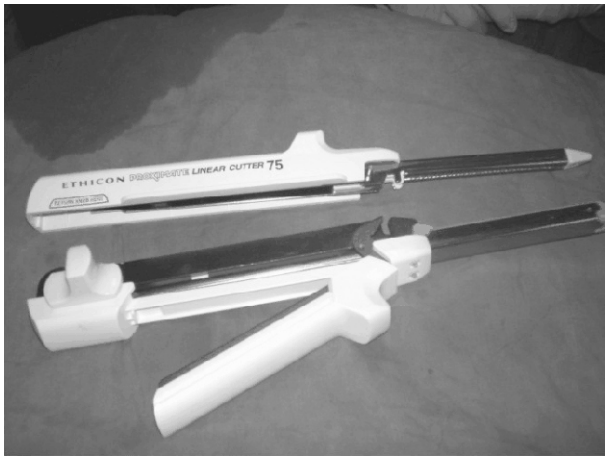
- Patients with sigmoid colostomy due to rectosigmoid aganglionosis proven by histopathology.
- Age more than 10 months.
- Weight more than 10 kg.

EXCLUSION CRITERIA

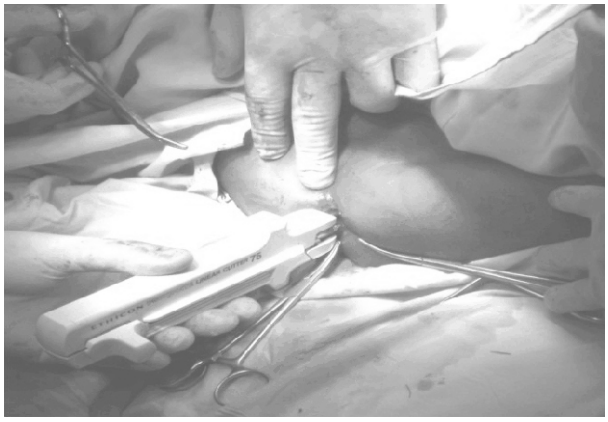
- Patients suffering from aganglionosis other than rectosigmoid region.
- Patients suffering from associated anomalies like trizomy 21.

PATIENTS AND METHOD

This was an interventional study conducted at Sheikh Zayed Hospital Rahim Yar Khan. The main aim, to conduct this study was to see the feasibility of Modified Duhamel procedure with linear cutter stapler device in a newly established Department of Paediatric Surgery at Sheikh Zayed Hospital Rahim Yar Khan. Proximate linear cutter was made available by communication with Ethicon Endo Company.(Photograph # 01)



Proximate linear cutter. Anvil half (upper) cartridge half (lower)



Proximate linear cutter. Anvil half (upper) cartridge half (lower)

Total seventeen patients from both sexes having sigmoid colostomy due to rectosigmoid aganglionosis were admitted through out patient department. Rectal biopsy confirmed Hirschsprung's disease and them sigmoid colostomy was performed at initial presentation. Patients suffering from aganglionosis other than rectosigmoid region or suffering from associated anomalies like trizomy 21 were excluded from the study. All routine investigations were done and it was confirmed that weight was more than 10 kg and Hb more than 10gm %.

Gut preparation was started before operation. Fresh blood was arranged and preoperative antibiotic was given. After general anesthesia with endotracheal intubation whole of the patient's abdomen and perineum was sterilized with pyodine. Elliptical incision was made around the colostomy site, proximal and distal loops were identified. Proximal loop (ganglionated colon) was mobilized so that it can reach the perineum without tension. Then retro - rectal tunnel was created and incision was made 1 cm above the dentate line in posterior half of anal canal. The ganglionic colon was brought up to this incision in pre sacral space and anastomosis was made.

The common wall of the aganglionic rectum and ganglionic colon also called "spur" was the main source of different modifications in classical Duhamel procedure. We used 75 mm proximate linear cutter stapler to cut and anastomose the spur, instead of old crushing technique. The upper end anastomosis was completed between rectum and colon. This completely renders two bowels into one lumen and obviates the possibility of a residual rectal pouch formation. Abdomen was closed after drainage with different set of instruments. After operation patient was shifted to the ward and kept nothing per oral till gut motility came.

Early complications like fever, vomiting, bleeding per rectum, retention urine, any wound infection, abdominal distension, anastomotic leakage and anastomotic stricture were noted and compared with other studies.

Monthly follow up record													
Parameters To Be noted		Visiting Months											
		1st	2nd	3rd	4th	5th	6th	7th	8th	9th	10th	11th	12th
Normal Stool evacuation	Once a day or more												
	Three per week												
	< three per week												
Abdominal Distension	Never in a month												
	Occasionally in month												
	Continuos												
Soiling	None in a month												
	< three per week												
	> three per week												
Stool Incontinence	Never												
	Daily												

Four parameters were noted and monthly follow-up record was maintained, according to following Performa

Scoring was done for following four parameters and then functional assessment was made as under;

1. Normal stool evacuation score

a. Once a day or more 1.0
b. Three per week 0.5
c. Less than three per week 0.0

2. Abdominal Distension

a. Never 1.0
b. Occasionally 0.5
c. Continuous 0.0

3. Soiling

a. None 2.0
b. Less than three per week 1.0
c. Three or more per week 0.0

4. Stool Incontinence

a. Never 1.0
b. Daily 0.0

TOTAL SCORE 0 – 5

FUNCTIONAL ASSESSMENT

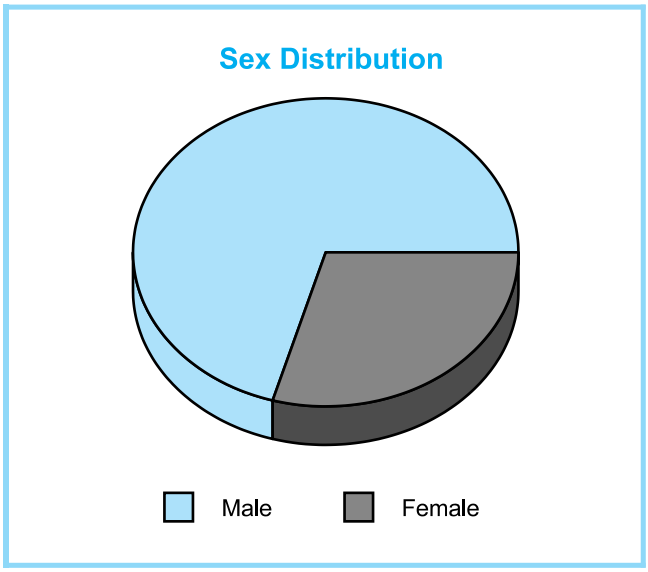
- Good functional results 4 – 5 points
- Fair result 2 – 3.5 points
- Poor results 0 – 1.5 points

RESULTS

Out of seventeen patients undergoing for modified duhamel procedure, twelve (70.59%) were male and five patients (29.41%) were female (Table-I Sex Distribution).

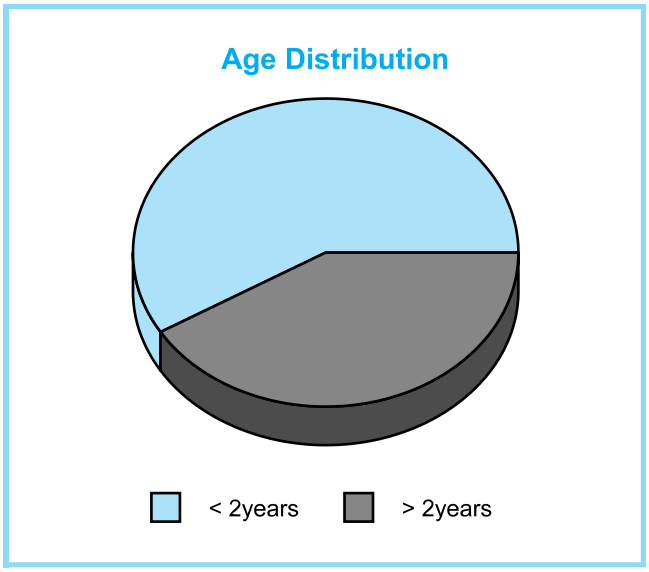
Table-I. Sex Distribution

Sex	Number	%age
Male	12	70.59%
Female	05	29.41%



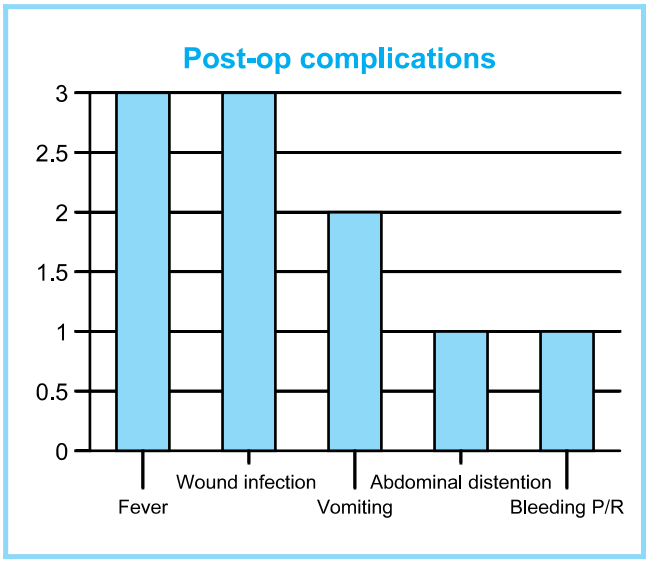
• Ten patients operated for Duhamel Procedure were less than 2 years of age (58.82%) and remaining 7 patients presented after two years (41.17%) (Table-II, Age distribution).

Table-II. Age Distribution		
Age	**Number**	**%age**
< 2 years	10	58.83
> 2 years	07	41.17



All seventeen patients were observed for post operative early complications. Out of them three patients (17.64 %) suffered from fever which was relieved by giving antipyretics and doing cold sponging. Three patients (17.64%) suffered from wound infection in which skin stitches were removed and daily dressing was done. Vomiting was observed in only two patients (11.76 %) who were managed conservatively. One patient (5.89%) suffered from abdominal distension on 3rd post operative day due to electrolyte imbalance and was relieved conservatively. There was mild bleeding per rectum in a girl (5.9%) and that was managed conservatively by giving vitamin K (Table-III, post-op complication).

Table-III. Post-op Complication		
Complication	**No. of Patients**	**% age**
Fever	3	17.60
Wound Infection	3	17.60
Vomiting	2	11.76
Abdominal Distention	1	5.89
Bleeding P/R	1	5.89

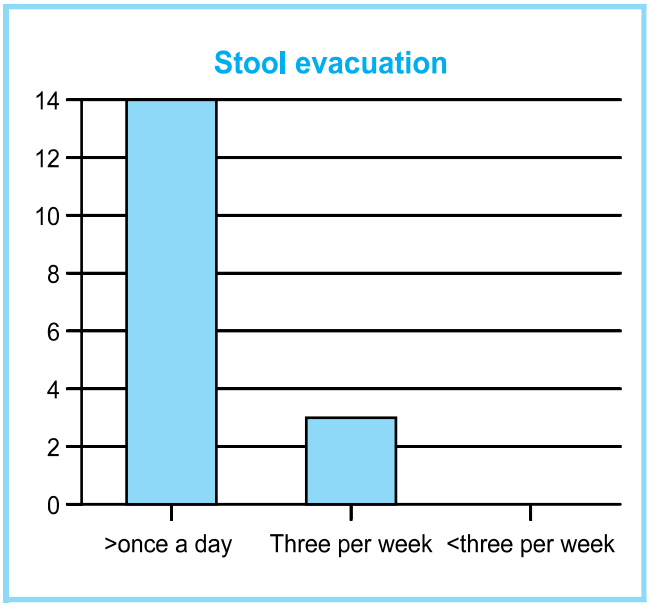


No patient suffered from retention urine, anastomotic leakage or anastomotic stricture.

Stool evacuation was the first important parameter noted in monthly follow up. Out of seventeen, 14 (82.35%) patients used to pass stool more than once per day. Initially stool evacuation was very frequent about 6-7 times a day but gradually it reduced up to 2-3 times. Only three patients (17.65 %) used to pass stool three times per week at the end of one year. No child passed stool less than three per week. (Table-IV, Stool Evacuation).

Table-IV. Stool Evacuation

Parameter	Patients	% age
> once a day	14	82.35
Three per week	03	17.65
< three per week	0	0



Abdominal distension was occasionally seen in 05 patients (29.41 %). In one patient distension was due to intentional obstruction and that was re-explored. In twelve patients (70.59 %) there was no distension after one year monthly follow up. No child suffered from continuous distension. (Table-V, Abdominal Distension).

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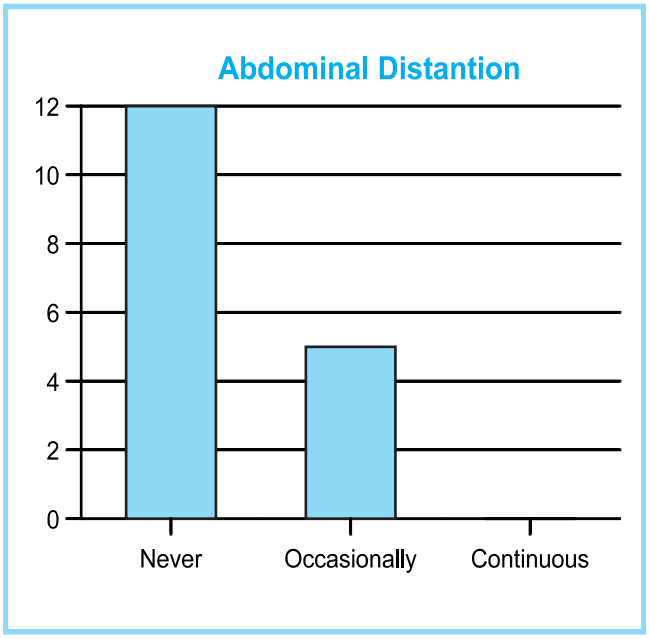
Table-V. Abdominal Distension

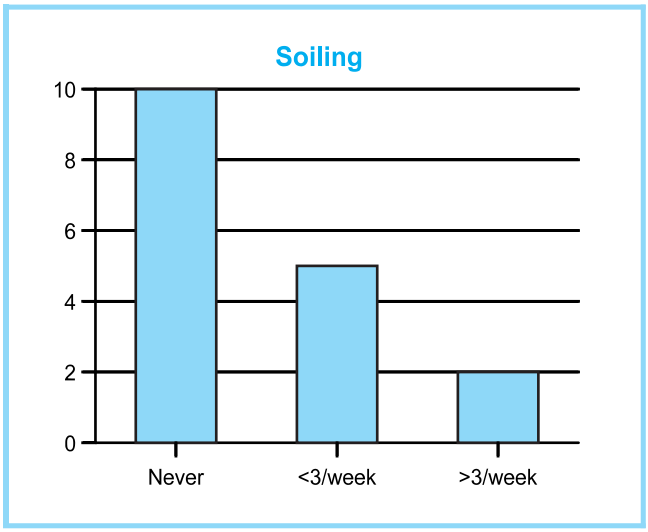
Parameter	Patients	% age
Never	12	70.59
Occasionally	05	29.41
Continuous	0	0

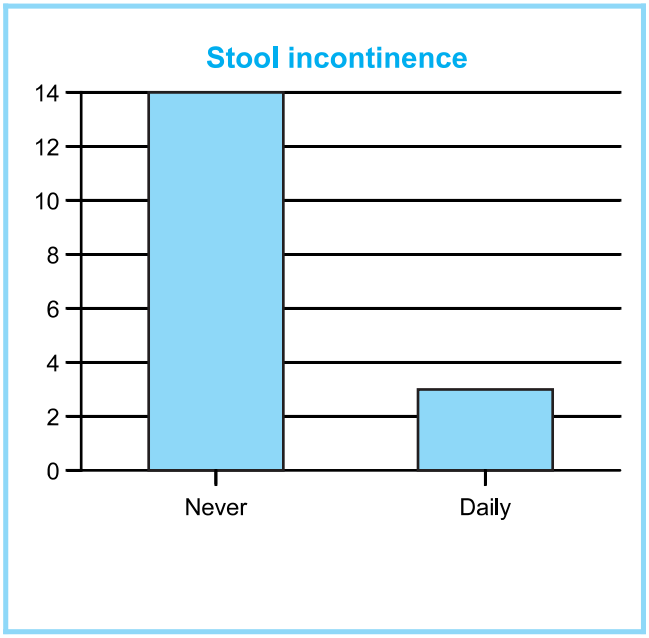


In ten (58.82 %) patients there was no soiling through out the year. Five patients (29.41 %) suffered from soiling for less than three times per week and in remaining two patients (11.76 %) soiling was frequent and they developed perianal excoriation. (Table-VI, Soiling).

Table-VI. Soiling

Parameter	Patients	%age
Never	10	58.83
< 3/ week	05	29.41
> 3/ week	02	11.76





Out of seventeen patients, fourteen patients (82.35 %) never developed stool incontinence. Three patients (17.64 %) developed incontinence and there was gradual improvement. (Table-VII, Stool Incontinence).

Table-VII. Stool Incontinence		
Parameter	**Patients**	**% age**
Never	14	82.35
Daily	03	17.65

Two patients developed enterocolitis 3rd and 7th months after surgery. Both cases were improved by giving antibiotics. There was no mortality during this study. After monthly follow up qualitative assessment was done by using scoring system as shown in table 8. (Table-VIII, Qualitative Assessment).

Following were the functional results after Modified Duhamel Procedure.

DISCUSSION

The Duhamel Procedure is widely used for definitive treatment of Hirschsprungs disease. The original technique which used two crushing Kocher Clamps as proposed by Duhamel in 1956, is no longer is use. Recently technical modifications using linear cutter stapler device have been introduced[17]. The procedure includes the use of a formal upper and lower anastomosis and division of spur by the GIA stapling device[18].

The results of our study are comparable with other national and international studies. In our study Modified Duhamel Procedure was performed in 17 patients. Out of them 05 patients (29.41 %) were females. According to Orr JD[19] male to female ratio in classic Hirschsprung Disease is 4:1 in favour of males. So in both studies males are affected more than females.

Table-IX. Functional Results			
Type of Results	**Points**	**No. of Pts.**	**%age**
Good Functional Results	4 - 5	12	70.59
Fair Results	2 - 3.5	5	29.41
Poor Results	0 - 1.5	Nil	Nil

Yanagihara j et al[20], performed modified Duhamel Procedure in 36 patients with GIA stapler and his results were very similar to our series. Six (16.6 %) of their patients developed enterocollitis while anastomotic leakage or stricture were not observed in any.

Occasional staining was observed in 05 patients (29.41 %) in our study. Bjornland K et al[21] conducted a study in

Table-VIII. Qualitative Assessment					
Patients #	Normal Stool Evacuation	Abdominal Distention	Soiling	Stool Incontinence	Scoring
1.	1.0	1.0	1.0	1.0	4
2.	1.0	1.0	2.0	1.0	5
3.	1.0	0.5	2.0	1.0	4.5
4.	0.5	0.5	1.0	1.0	3
5.	1.0	1.0	2.0	1.0	5
6.	1.0	1.0	2.0	1.0	5
7.	1.0	1.0	0.0	1.0	3
8.	0.5	1.0	2.0	1.0	4.5
9.	1.0	0.5	1.0	1.0	3.5
10.	1.0	1.0	2.0	1.0	5
11.	1.0	1.0	2.0	1.0	5
12.	1.0	1.0	1.0	1.0	4
13.	1.0	1.0	1.0	1.0	4
14.	1.0	0.5	1.0	1.0	3.5
15.	1.0	1.0	2.0	1.0	5
16.	1.0	1.0	0.0	1.0	3
17.	1.0	1.0	2.0	0.0	4

1998 in which 48 patients were operated. In his study occasional soiling was 31.3% and normal fecal control was 60.40% while in our study normal fecal control was 82.35%. In another study conducted by Marty et al[22] in 1995 normal fecal control was 65 %. So fecal control is quite satisfactory in our study.

Modified Duhamel Procedure for Hirschsprung Disease with the help of stapling device is a safe and easy procedure with minimum morbidity and no mortality. Complications like mild enterocolitis, mild constipation or soiling can be dealt conservatively. Same results were drawn by Mottioli G et al[23] in1998.

CONCLUSION

After comparison with other international studies it is concluded that Modified Duhamel Procedure for Hirschsprung's Disease with the help of Mechanical Stapling Device is quite safe, easy and less time consuming. It has got relatively good results as compared to other procedures. That is why Modified Duhamel with the help of staplers is the procedure of choice in Pediatric Surgery Department, Sheikh Zayed Hospital, Rahim Yar Khan. It can be adopted confidently for Hirschsprung's disease. However there is need to conduct this study on large series of patients.

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