ORIGINAL ARTICLE

Hirshsprung disease: Shall we move forward? A retrospective cohort study

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Abstract:
Objective: To determine the safety and effectiveness of singles stage corrective surgery in Hirshsprung disease (HD).
Place and duration of the study: Liaquat National Hospital and Medical College, Department of Pediatric Surgery, done from January 2007 to January 2020.
Material and Methods: A retrospective review of data was of all the cases operated for biopsy proven Hirshsprung disease that had not undergone any definitive corrective procedure. Record retrieval was done though outpatient patient registry via file numbers. An informed consent was taken from the parents and all children were divided into 2 groups, GROUP 2 (STAGED ERPT) included children who had under gone staged repair while GROUP 1 (primary endo-rectal pull through) included children who had a single stage endorectal pull through (ERPT) corrective procedure. Primary outcomes were total hospital stay and quality of life scoring on follow-up. Secondary outcome variables included immediate post-procedure complications such as surgical site infection (SSI), anastomotic dehiscence, anastomotic stricture formation, peri-anal excoriation and post-operative ileus. Post-operative morbidity was assessed using Clavien-Dindo classification. Children who were unable to come for follow-up were consulted by telephonic interview, and Performa was filled.
Results: A total of 47 patients were operated for Hirshsprung disease between January 2007 and June 2020. 18 children underwent primary single stage endo-rectal pull through, while 29 patients underwent a staged endo-rectal pull through procedure. Both groups had male predominant sex distribution, Group 1 (n=18) there were 15 male children and 3 female children. Group 2 (n=29) had 23 male children and 6 female children. Children in group 1 had mean age of diagnosis 3 months, and underwent primary endo-rectal pull through at mean age of 5 months. Their weight at the time of definitive procedure was a mean of 5.7kg. Children in group 2 had mean age of diagnosis 21 months, and underwent pull through procedure at mean age of 26 months. Their weight at the time of definitive procedure was 9.0 kg. A comparative review revealed that patient who underwent primary endorectal pull through had a shorter mean hospital stay of 5.3 days as compare to patients who underwent a staged ERPT, mean 10.5 days of hospital stay (p value 0.047). we used quality of life scoring scale evaluating fecal soiling, stooling frequency and need for diapers in post-operative follow-up period there was no statiscally significant diffenrece in between the two groups (p value 0.090). There were clearly greater post operative complications and admissions in patients who underwent staged ERPT. 17.24 % of children who under-went staged ERPT developed SSI in comparison to none in Primary ERPT group. (p value 0.045). Incidence of post-operative ileus was also higher in Staged ERPT (20.96%) group as compare to Primary ERPT group (5.56%). However it was statistically not significant. Rests of the post-operative complication were comparable in the two groups. Post-operative enterocolitis was the most common cause of hospital re-admission post-operatively and the incidence was comparable in between the 2 groups. Over all morbidity post procedure was Grade 1 as per Clavien Dindo classification system. 2 patients in Staged EPRT group required re operation hence falling in GRADE 3 of ClavienDindo grading system, but was satistically not significant (p value 0.08).
Conclusion: Primary endorectal pull through is a safe procedure for patients with Hirshsprung disease as there is avoidance of stoma formation, there is better continence and low incidence of post-operative complications.
Keywords: Primary endorectal pull thorugh (ERPT), trans-anal, Hirshpsrung disease (HD), staged ERPT, Clavien Dindo classification

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Introduction:

The hirshpsrung's disease defines as congenital a-ganglinosis of a segment of large bowel was first described in 1988 by a Danish pediatrician

as a chronic form of severe constipation that resulted in a congenital megacolon. The incidence has been estimated to be 1 in 5000 live births. A variety of procedures have been attempted. The objective of the surgical treatment is to resect the a-ganglionic segment of the bowel and re-establish the continuity of ganglionic bowel with anal canal.

The traditional management is two or three staged surgical procedure. After confirming the diagnosis by rectal biopsy, leveling colostomy is done. Few weeks or months after this aganglionic bowel are excised and continuity reestablished with or without a covering stoma. Soave endo-rectal pull through was introduced by Franco Soave at Institute G. Gaslini in 1985. Greater awareness of Hirshsprung disease improved nursing care, use of suction rectal biopsy and pediatric pathologist have all contributed to earlier diagnosis and management for these infants. The use of a primary endorectal pull through in the surgical management of patients with Hirshsprung disease represents a remarkable change from the classic approach.

Staged procedure is known to associate with grave discomfort to the child and parents, besides associated with morbidity and increases the cost of treatment. Acceptance of stoma also cultures into an issue. Since 1988, Dela Tore-Mondragon and Ortege first describes single stage transanal pull though, many specialized pediatric surgery centers have taken up the single stage procedure with different modifications with time. The single stage has been performed by open technique, with laparoscopic assistance and using purely transanal approach. It has led to decrease in operative time, total hospital stay and avoidance of stoma. In view we at Liaquat National Hospital started performing single stage procedure. Aim of this retrospective study is to evaluate safety and effectiveness of single stage procedure.

Material and Methods:
A review of data was done from January 2017 to January 2020 of all the cases operated for biopsy proven Hirshsprung disease that had not undergone any definitive corrective procedure. Children who had undergone a previous corrective procedure and those with incomplete data were excluded from the study. Record retrieval was done though out-patient patient registry via file numbers. Hospital review board permission as taken. All parents were informed, counselled and consented for review and questionnaire. GROUP 2 (STAGED ERPT) included children who had under gone staged repair while GROUP 1 (PRIMARY ERPT) included children who had a single stage endorectal pull through (ERPT) corrective procedure. All patients were followed post-operatively. Primary outcomes were total hospital stay and quality of life scoring on follow-up. Secondary outcome variables included immediate post-procedure complications such as surgical site infection, anastomotic dehiscence, anastomotic stricture formation, peri-anal excoriation and post-operative ileus. Post-operative morbidity was assessed using Clavien-Dindo classification. Children who were unable to come for follow-up were consulted by telephonic interview, and performa was filled. All data are presented as mean. The Mann–Whitney U test was used to compare outcomes between groups with non-parametric data. The Chi-Square test was used to compare outcomes between categorical data. $P<0.05$ was considered statistically significant. Operative definition for stages repair, was children undergoing definitive procedure after previous stoma. Total hospital stay was defined as sum of total days of stay when patient was admitted for diagnosis plus definitive procedure. IBM SPSS version 22 used for statistical analysis of data.

Results:
A total of 47 patients were operated for Hirshsprung disease between January 2017 and June 2020. 18-children underwent primary single stage endorectal pull through, while 29 patients underwent a staged endorectal pull through procedure. Both groups had male predominant sex distribution, Group 1 (n=18) there were 15 male children and 3 female children. Group 2 (n=29) had 23 male children and 6 female children. Children in group 1 had mean age of diagnosis 3

Table 1: Patient Characteristics

	Single stage ERPT n=18	Staged procedure n=29
Median age at presentation	5 months Range (3rd day of life till 15 months)	21months Range (5th day of life till 36 months)
Mean age at definitive surgery	5 months (1 month – 9 months)	26 months (6 months – 36 months)
Sex		
Male	15	23
Female	3	6
Mean weight at definitive procedure	5.7 kg (4.3 kg- 8.0kg)	9 kg (5kg – 15kg)

Table 2: Primary outcome measure

	Single stage ERPT n=18	Staged Procedure n=29	p value
Total hospital stay (mean)	4.5 days (3.5 – 6 days)	7 days (5 – 9 days)	0.170
Quality of life scoring Fecal soiling Stooling frequency Need for diaper	7/10	5/10	0.090

Table 3: Complications post-operative after ERPT

	Single Stage ERPT n=18	Staged procedure n=29	p value
Surgical site infection	0	5 (17.24 %)	0.045
Anastamotic dehiscence	0	0	-
Anastamotic stricture	0	1 (3.44%)	0.080
Perianal excoriation	3 (16.67%)	6 (20.70%)	0.190
Ileus	1 (5.56%)	6 (20.68 %)	0.054

Table 4: Complications immediate post-operative after ERPT

CLAVIN-DINDO classification	Single stage ERPT n=18	Staged procedure n=29	p value
Grade 1	3 (16.67%)	7 (24.71%)	0.27
Grade 2	1 (5.56%)	2 (6.90%)	0.11
Grade 3	0	2 (6.90%)	0.08
Grade 4	0	0	

Table 5: Re-Admission due to hd related problems post-operatively

	Single Stage ERPT n=18	Staged Procedure n=29	p value
Enterocolitis n(%)	2 (11.11%)	3 (10.34%)	0.320
Intestinal obstruction n(%)	0	2 (7.0%)	0.100

months, and underwent primary ERPT at mean age of 5 months. Their weight at the time of definitive procedure was a mean of 5.7kg. Children in group 2 had mean age of diagnosis 18 months, and underwent pull through procedure at mean age of 20-months. Their weight at the time of definitive procedure was 9.0 kg as shown in table-1. A retrospective comparative review revealed that patient who underwent Primary ERPT had a shorter mean hospital stay following definitive procedure of 5.3 days as compare to patients who underwent a staged ERPT, mean 10.5 days of hospital stay (p value 0.047). We used a qualitative quality of life scoring scale evaluating fecal soiling, stooling frequency and need for diapers in post-operative follow-up period. For patients who underwent primary ERPT is was good grading 7/10 on average, while patients who underwent a staged primary procedure had fair grading 5/10 score on average. (p value 0.090) as shown in table-2

Post-operative complications which included SSI, anastomotic dehiscence, anastomotic stricture, peri-anal excoriation and ileus were assessed and Hirshsprung disease associated causes of re-admission post-operatively specifically enterocolitis and intestinal obstruction were analyzed. There were clearly greater post-operative complications and admissions in patients who underwent staged endo-rectal pull through, as pointed out in table-3. 17.24% of children who underwent staged endo-rectal pull through developed SSI in comparison to none in primary endo-rectal pull through group. (pvalue 0.045). Incidence of post-operative ileus was also higher in staged endo-rectal pull through (20.96%) group as compare to primary end-orectal pull through group (5.56%). However it was statistically no significant. Rests of the post-operative complication were comparable in the two groups. Post-operative enterocolitis was the most common cause of hospital re-admission post-operatively and the incidence was comparable in between the 2 groups as shown in table-5).

We classified overall progress of all patients and need for intervention as per Calvin Dindo classification system defining the safety of procedure in both groups as shown in table-4. None of the patients went into multi-system organ

failure or developed mortality in both groups of patients. Over all grades was Grade-1 as per clavien Dindo classification system. 2-patients in Staged EPRT group required re operation hence falling in GRADE 3 of Clavien Dindo grading system, but was satistically not significant (p value 0.08).

Discussion:

Hirschsprung's disease is a common surgical problem in children. It is characterized by absence of ganglion cells in mesenteric and myenteric plexus of distal bowel extending to variable level proximally.[12] In majority of the children, the diseased segment extends up to the recto-sigmoid junction. First definitive surgical procedure was described by Swenson. Previously, multi-stage surgery was the preferred mode of management.[9] Over the past two decades, it has been increasingly recognized that the routine use of a colostomy is unnecessary and an increasing number of pediatric surgeons perform the reconstruction as a single stage procedure at an early age. The single stage management of Hirschsprung's disease avoids colostomy and its related complications including the social stigma of a stoma.[11] The short operating time and hospital stay with encouraging early results at affordable cost makes it the obvious choice over traditional multistage procedures.[5]

We at Liaquat National Hospital started doing single stage transanal procedure in 2017. Since then all patients with Hirschsprung's disease presented primarily to us and are in good nutritional and physiologic state undergone trans-anal-endorectal pull through. Patients presenting with stoma, operated primarily else where or those who were thought not suitable for single stage procedure because of the state of bowel or nutrition, undergone staged procedure and acted as comparable group. In our study the average age of primary corrective single stage procedure was 5-months. The post-operative hospital stay was lower in patient swho underwent Single stage endorectal pull through compared to comparative group, this was possibility due to longer period of post-operative ileus. One of the major issues in the trans-anal approach is the significant stretching of the anal sphincters during surgery with its potential impact on continence in later life. However, this is only transient and bowel movements became normal in majority of cases within a period of 2 weeks to 3 months. In our study, the quality of life scoring post-surgery in the two groups were comparable as meticulous protection of the anal canal and sphincter complex was done during corrective endo-rectal pull through. Entero-colitis has been considered as one of the main problems in patients with Hirschsprung's disease both before and after definite treatment. The incidence of post pull-through entero-colitis reported in the literature varies widely with some studies reporting rates as high as 32 to 42%. Hackman et al[4] studied the risk factors for post-operative entero-colitis and found that both the presence of anastomotic leak or stricture and the development of post-operative intestinal obstruction secondary to adhesions increased the relative risk and subsequent entero-colitis by approximately 3 fold. Long seromuscular cuff and a high colo-anal anastomosis are associated with increased risk of post-operative enterocolitis. In this study, 5-patient developed enterocolitis in the follow-up period, more in staged endorectal pull through group and was managed with intravenous fluids, rectal wash and intravenous antibiotics.

It was highlighted that majority of the children had grade-1 morbidity as per Clavien Dindo classification. However due to greater number of surgical procedure overall in children in staged group had higher risk of post-operative adhesions and went into adhesive obstruction requiring operative intervention falling in grade 3, improving the safety profile of primary endorectal pull through.

As far as limitations of the study are concerned it is a single center study hence results are not generalisable. Patients, who did not have an out-patient follow-up, were interviewed on telephone which can be subject to recall bias. The sample size is small, a larger multi-center study is required to effectively identify the safety profile of the conventional staged endo-rectal pull

through and primary endorectal pull through and may produce statistically significant results.

Conclusion:
Primary endo-rectal pull through is a safe procedure for patients with Hirshsprung disease as there is avoidance of stoma formation, there is better quality of life scoring and low incidence of post-operative morbidity.

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Role and contribution of authors:
Tahir Muhammad Yaseen, collected the data, references and did the initial writeup.

Shabbir Hussain, critically review the article and made final changes.

Taimoor, collected the data, and helped in introduction and discussion writing.

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