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Single Stage TransanalEndorectal Pull-Through(T.E.P.T) For Hirschsprung'sDisease

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Abstract*:Hirschsprung's disease is a common cause of intestinal obstruction in pediatric age group. It usually present in the neonatal period or early childhood. Diagnosis is usually made by classical presentation of H.D, supported by barium enema showing a transition zone. Rectal biopsy showing absent ganglion cells, nerve hypertrophy and increased acetyl cholinesterase on immune-histiochemical study is confirmatory. Diverting colostomy is done at the time of diagnosis, definitive surgery is performed when the child attains sufficient weight and age followed by closure of colostomy. The three stages procedures were subsequently converted to a two stage procedures by foregoing a protective colostomy for the definitive surgery. The disadvantages of staged procedures, like multiple surgeries, repeated hospitalization and colostomy management and its complication leads to the development of single stage procedure for hirschsprung'sdisease.Primarytransanal approach apart from being single stage, has added advantages of no peritoneal breach, no injury to pelvic innervations or other pelvic organs, better cosmesis, no abdominal scar, less hospital stay, cost effective, less post-operative pain and practically no risk of developing adhesives obstruction. Our study is a retrospective as well as prospective study of initial 30 consecutive cases of hirschsprung's disease who underwent single stage transanal pull through in the department of pediatric surgery at Coimbatore medical college hospital, Coimbatore from Jan 2015 to Jan 2018. The results of the study shows low perioperative complications, and out comes comparable with various studies ,and also reduces the number of surgeries needed. Thus this technique with its all advantages has a role in surgical management of Hirschsprung's disease especially in neonates.*

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I. Introduction

Hirschsprung's disease (HD), named after HaraldHirschsprung, Who presented the classic description in 1886, is a common cause of intestinal obstruction in pediatric age group. The incidence ranges from 1 in 4500 to 1 in 7000 live births, with 4:1 male to female ratio.[5,6]It usually present in the neonatal period or early childhood. Atypical presentation in adolescents in the neonatal period or early childhood. Atypical presentation in adolescents and adults is not uncommon. Neonates usually present with delayed passage of meconium beyond 48 hours, abdominal distension and vomiting. Beyond neonatal period the presentation is that of chronic constipation, abdominal distension and failure to thrive.

The exact etiology is not known, but the basic pathology lies in the failure or blockage of migration of neuroenteric cells from the neural crest to the alimentary tract. This results in absence of ganglion cells both in Meissner's and auerbach's plexus of GIT. Moreover there is an increase in the aganglionic segment. This leads to spastic, non- relaxing, non-peristaltic aganglionic segment causing obstruction.

Diagnosis is usually made by classical presentation of H.D, supported by barium enema showing a transition zone. Rectal biopsy showing absent ganglion cells, nerve hypertrophy and increased acetyl cholinesterase on immuno-histochemical study is confirmatory.

Over the last century the therapeutic options for hirschsprung's Disease have gradually undergone refinement through trial and error. First treatment was a diverting colostomy but the symptoms recurred after closure of colostomy. Subsequent attempts at bypass or removal of redundant portion of colon were uniformly unsuccessful. Ladd and gross in 1941 reported improvements in symptoms after lumbar sympathectomy.[7,8,9] First successful surgical technique described by Bernard Duhamel in 1956[14] and endorectal pull through technique described by Franco soave in1960 and subsequently modifies by Boley[16,17], gained worldwide popularity.

All these techniques namely Swenson's, Duhamel's or soave's are usually two staged or even three staged procedures. Diverting colostomy is done at the time of diagnosis, definitive surgery is performed when

the child attains sufficient weight and age followed by closure of colostomy. The three stages procedures were subsequently converted to a two stage procedures by foregoing a protective colostomy for the definitive surgery.

The disadvantages of staged procedures, like multiple surgeries, repeated hospitalization and colostomy management and its complication leads to the development of single stage procedure for hirschsprung's disease. The initial procedures(1980) were primary endorectal pull-through without preliminary colostomy done by abdominal approach. Further development was in the form of laparoscopic- assisted primary pull-through described in1995.

Primary transanalendorectalpull-through was first described in1998.[20]Transanal approach apart from being single stage, has added advantages of no peritoneal breach, no injury to pelvic innervations or other pelvic organs, better cosmesis, no abdominal scar, less hospital stay, cost effective, less post-operative pain and practically no risk of developing adhesives obstruction.

The purpose of a pull through procedure for hirschsprung's disease is to remove the aganglionic colon, bring normally innervated bowel to the anus, and preserve anal sphincter function. Although all the three commonly performed procedure accomplish these goals, the operation described by soave and adapted by boley has the advantage of avoiding the retrorectal plane, thereby eliminating the possibility of injury to the pelvic nerves responsible for urinary continence and sexual function.

The use of laparoscopy to perform pull-through surgery for HD has been growing in popularity in recent years. All the three standard procedure have been adapted to minimally invasive approach. All use at least three abdominal port sites with the associated risk of bleeding and thermal or harmonic injury to other pelvic structures. Laparoscopic surgery requires adequate expertise, experience and assistance. Single stage transanalendorectal pull through provides the same advantages as laparoscopic surgery with additional advantage of eliminating risk of intraabdominal pelvic dissection and adhesion formation, no scar and less pain.

Various studies over last few years have reported the results of transanalendorectal pull-through as good as, if not better than staged procedure. Single stage transanal pull through was started in our institute in year 2012. This study was undertaken to evaluate the results of this procedure in the initial 30 cases performed, as not much has been reported from our country. This study is an attempt to evaluate the early results of this new but promising technique of primary transanalendorectal pull-through for rectosigmoidhirschsprung's disease.

II. Materials And Methods

Inclusion criteria:

The following patients were included in the study..

1 Only classical type of hirschsprung's disease.
2 No preliminary colostomy.
3 Pateints with no evidence of perforation or enterocolitis at diagnosis.

Exclusion criteria:

The following patients were not included in the study.

1 Patients with ultra-short segment disease, long segment disease and total colonic aganglionosis.
2 Patients with prior colostomy.
3 Patient with evidence of perforation or enterocolitis.
4 Patients with obstruction not relieved by rectal irrigation.
5 Associated life threatening anomalies.
6 Patients with more than 6yrs of age.

This is retrospective as well as prospective study of initial 30 consecutive cases of hirschsprung's disease who underwent single stage transanal pull through in the department of pediatric surgery at Coimbatore medical college hospital, Coimbatore from Jan 2015 to Jan 2018.

All patient with history suggestion of hirschsprung's disease were subjected to barium or contrast enema in the radiology department of the institute. Contrast enema was postponed 24 to 48 hours beyond any kind of rectal manipulation like irrigation, suppository or digital examination. A catheter is placed just inside the anus and contrast is injected till dilated bowel is outlined. A well defined transition zone that is a contracted distal bowel was a prerequisite for diagnosis of hirschsprung's disease.

Patients who had their transition zone confined to rectosigmoid junction or distal to rectosigmoid were eligible for single stage transanalendo rectal pull-through procedure (TEPT). More proximally located transition zone, long segment hirschsprung's disease and total colonic aganglionosis were managed by staged procedure.

Patient with previous colostomy were not included in the study. Similarly patients presenting with bowel perforation, active enterocolitis or obstruction where bowel could not be successfully decompressed by rectal irrigation were considered not suitable for single stage transanalendorectal pull through.

Rectal biopsy was not done in any patient to establish the diagnosis of hirschsprung's disease as, firstly barium enema study showing a well defined transition was always diagnostic of hirschsprung's disease, secondly rectal biopsy would not have shown the level of aganglionosis prior to surgery, thirdly rectal biopsy would have made subsequent mucosal dissection difficult due to result of adhesion formation.

A data chart was designed to collect following data;

Patients demographics including sex,age,gestation,and weight at diagnosis

1 Preoperative clinical data (presenting signs and symptoms and diagnostic studies).
2 Operative details including type of pull through, length of bowel resected, intraoperative complications and blood transfusion requirement.
3 Early Postoperative course like degree of pain timing of starting oral feeds.
4 Early and late complications
5 Additional surgical procedures, if any.
6 Functional outcome as judged by bowel habits and anorectal continents.
7 Histopathological findings.

Data was collected retrospectively from hospital records and prospectively in some as and when they were admitted for pull through procedure. Intern follow up data was obtained at revisits. Final follow up data was compiled by personal interview of the parents and physical examinations of the patients.

Pre-operative preparation:

Once bowel decompression was ensured by frequent rectal irrigation mainly infants and older children, patients were planned for singe stage transanalendorectalpull through. The day prior to surgery every patient received total gut irrigation. Through a Naso-gastric tube normal saline at a rate of 20ml/kg/hr was infused along with intermittent rectal washes till the effluent per anus was clear and free of fecal residue. This usually took 4to 6 hours.

Surgical Technique:

After induction of anesthesia , the patient was placed in lithotomic position with pelvic elevated at the end of operative table. Intravenous anti-biotic, often a third generation cephalosporin was given at induction and bladder was catheterized by an inhaling catheter. Naso-gastric tube inserted for total gut irrigation was maintain.

Mini laparotomy done in left lilac fossa, transition zone identified,marker stich taken about 3-5 cm above transition zone at antimesentric border, in some cases we done laparoscopy to identify transition zone and to place marker stitch.

The anal canal was exposed by means of stay sutures. Submucosal injection of saline with epinephrine was used. A circumferential incision was made 0.5cm proximal to dentate line and mucosal sleeve was dissected and extended proximally.

Once the submucosal plane is established the dissection was continued proximally using blunt dissection and cauterization of submucosal infiltrating vessels. Traction on the mucosal tube facilitates proximal to peritoneal reflection.

Stay sutures were inserted to control the upper end of muscular cuff, which was incised circumferentially allowing exposure of full thickness sigmoid colon. Mobilization of colon is continued proximally by ligating and dividing the rectosigmoid vessels till the dilated portion of the colon or marker stitch is reached.

As a routine colon was usually mobilized 4-5cms proximal to the appearance of transition zone to avoid areas of hypoganglionosis adjacent to the aganglionic zone for coloanal anastomosis. After resection of the aganglionic segment the muscle cuff in incised longitudinally in the midline posteriorly under direct vision. Care was taken to avoid injuring the anal sphincter muscles distally.

The ganglionic bowel was then pulled through the muscular cuff and anastomosed to the rim of remaining mucosa above the dentate line with 4-0 polygalatin sutures.

Dressing was done with a Vaseline gauze pack in the anal canal. Anal pack and urinary catheter was removed on the first post-operative morning. Oral feeding was starting by second or third post operative day and gradually shifted to regular diet appropriate for age. Intravenous antibiotics were continued till the oral feeding was tolerated usually first 48 hours, following which patient shifted to oral antibiotics for the next 5 days. Post operative pain control was achieved by mild analgesics like paracetamol. Opioids or sedation was not required in any patient for the pain relief. caudal anesthesia block was given to all patients at induction and was very helpful in achieving pain free immediate post operative period.

Patients were discharged once they started tolerating complete oral diet and regular passage of stools was ensured. First follow up was visit ws usually 2 weeks after surgery. Later, regular followup was initially

monthly and then 3 to 6 monthly. Due to various social obligations and difficulties in our country fraction of patients did not adhere to the follow up routine.

Regular anal dilation was not carried out for initial few patients. Due to occurrence of anastomotic strictures in some of the initial patients, we restored to routine anal dilation starting one month after surgery.
No routine follow up investigations were done in our study.

III. Results

A total of 30 patients were includes in this study from January 2015 to January 2018. Of these patients 26 were male patients (86.6%) and 4 were female patient (13.3%).

Table 1: Age profiles of the patients

Percent Patient group	Number	Percentage
Neonates	10	33.3%
Infants	13	43.3%
children	7	23.3%

Majority were infants (43.3%) less than one year of age. Neonatal presentation was 33.3% of the cases. 23.3% were children above one year of age.

Table 2: Age and weight distribution

Percent Patient group	Mean age (Range)	Mean weight (Range)
Neonates	8.7 days(3-20)	2.87 Kg(2.5-3.3)
Infants	5.11 months (2-12)	5.63 Kg (2.7-9.5)
children	4.35 years (2.5-6)	12.62 Kg (9.2-16.0)

All the patient were term delivers with no specific perinatal complications.
All neonates were good weight, mean weight being 2.87Kg (2.5-3.3).

Table 3: presenting symptoms

Symptoms	Delayed passage of meconium	Distension	Vomiting	Chronic constipation	Chronic laxative use	Enterocolitis
Neonates (n=10)	10(100%)	10(100%)	4(40%)	2(20%)	0(0%)	0(0%)
Infants (n=13)	11(84.6%)	12(92.3%)	6(46.1%)	12(92.3%)	10(76.9%)	2(15.3%)
Children (n=7)	4(57%)	7(100%)	3(42%)	7(100%)	7(200%)	1(14.28%)
Total (n=30)	25(83.3%)	29(96.6%)	13(43.3%)	21(26%)	17(56%)	3(10%)

All neonates presented with the history of delayed passage of meconium and abdominal distension. Vomiting was present in 40% of the neonates. The commonest presenting symptoms in infants were abdominal distension and constipation (92.3%). All children above one year of age presented with abdominal distension and chronic constipation requiring regular use of laxatives or suppositories. Enterocolitis was present in only 10% of the cases.

Table 4: **Onset and duration of symptoms**

Group	Since birth	Delayed
Neonates (n=10)	10(100%)	0
Infants (n=13)	10(76.9%)	3(23.1%)
Children (n=7)	4(57)	2(28.5%)

All neonates and majority of infants were symptomatic since birth. Only half of the children groups were symptomatic since birth, rest had delayed onset of their symptoms.
The mean age at which symptomatology appeared is 3 years in children (1-5 years) and 3 months in infants (2-5 months) in whom onset was delayed.

Table 5: clinical examination findings

	Distention of abdomen	**Visible or palpable bowel loops**	**Visible peristalsis**	**Palpable fecalomas**	**Explosive passage of stools on per rectal examination**	**pallor**
Neonates (n=10)	10(100%)	5(50%)	3(30%)	0	5(50%)	0
Infants (n=13)	11(84.6%)	7(53.8%)	0	2(15.3%)	3(23.0%)	4(30.7%)
Children (n=7)	6(85.7%)	3(42.8%)	0	3(42.8%)	2(28.5%)	3(42%)
Total (n=30)	27(90%)	15(50%)	3(10%)	5(16.6%)	5(33.3%)	7(23.3%)

Abdominal distension was the commonest clinical finding in all age groups. Palpable or visible bowel was more commonly a feature in younger patients, whereas fecalomas were palpable more commonly in older children. Explosive passage of stools after digital rectal examination was more frequent in neonates compared to infants and children. About 30% of the infants and children had pallor at the time of presentation.

One patient had associated downs syndrome apart from which no other associated anomalies were present in our study group.

Investigative results

Renal function test and serum electrolytes were normal in all patients. Anemia (hemoglobin level of less than 10gm %) was seen in 4 of 13 infants (30.7%) and 3 out of the 7 children (42.8%).

Table 6: contrast enema findings

	Rectum	**Rectosigmoid**	**Sigmoid**
Neonates (n=10)	4(40%)	6(60%)	---
Infants (n=13)	3(23.1%)	9(69.2%)	1(7.6%)
Children (n=7)	3(42.8%)	4(57%)	---
Total (n=30)	10(33.33%)	19(63.33%)	1(3.3%)

63% of the contrast enema study revealed a transition zone at recto sigmoid junction, while transition zone confined to rectum was seem in 33.33% of the cases. Transition zone at the level of sigmoid was noted in one patient. A well-defined transition zone could be identified in all patients.

Table 7: Length of intestine resected

	Length of resected bowel(in cms)
Neonates (n=10)	10.4(7-16)
Infants (n=13)	12.8(8-20)
Children (n=7)	12.9(6-15)
Total (n=30)	12.3(6-20)

Mean length of bowel resected was more in older children compared to neonates.

Table 8: Requirement of blood transfusion

	Number of patients who received transfusion
Neonates (n=10)	4(40%)
Infants (n=13)	7(53.8%)
Children (n=7)	5(71.4%)
Total (n=30)	16(53.33%)

Blood transfusion requirement was higher in children above one year compared to neonates and infants. 53% of the total patients required blood transfusion during or after surgery.

The paraffin section histopathology report was consistent with diagnosis of hirschsprung's disease in 25/30 patients (83.33%). Of these 25 patients 3 patients (10%) had absent ganglion cells at the proximal limit of the resection. All 3 had well defined transition zone on contrast enema. Of this three patient one patient had continues obstructive symptoms in the post operative period. Repeat contrast enema of this child revealed persistent transition zone and this child further underwent a one stage Duhamel's procedure. The 2nd patient in this category is having normal bowl function at 9 months of follow up. The 3rd patient has been lost to follow up.

In five patient the paraffin section showed ganglion cells through out the length of the resected specimen. All of these patients however showed well defined transition zones on their respective contrast enemas. Out of this five patients three patients have normal bowel habits while one is suffering from recurrent attacks of enterocolitis and constipation. The last patient in this category had expired 15 days after surgery.

The possible explanation for presence of ganglion cells along whole length of resected specimen while contrast enema revealed a well defined transition zone is that, patient with transition zone confined to rectum or a short segment of aganglionosis might have been missed in paraffin section study of the mucosectomy specimen.

Table 9: Timing starting post-operative oral feeds

	Mean day of starting oral feeds (range)
Neonates (n=10)	2.7 days (2-4)
Infants (n=13)	2.4 days (2-3 days)
Children (n=7)	2.1 days(1-3 days)
Total (n=30)	2.4 days (1-days)

Early oral feeding post-operatively was feasible in all age groups. Older children tolerated oral feeds earlier compared to neonates in the post-operative period.

Table 10: duration of hospital stay

	Rectum	**Rectosigmoid**
Neonates (n=10)	12.4 days (9-17 days)	5.1 days (4-7 days)
Infants (n=13)	9.23 days(5-23 days)	4.1 days (3-6 days)
Children (n=7)	11.7 days (5-26 days)	4.7 days (3-7 days)
Total (n=30)	10.8 days (5-26 days)	4.5 days(3-7 days)

All patients were discharged within a week after surgery (mean 4.5 days). Pre operative hospitals stay was determined by the amount of time taken for complete bowel decompression by means of rectal and colonic washes.

Follow up period

Means follow up period is 17.3 months range being 5 months to 38 months. Of the total patients in the study group, 8 patients were lost to follow up. Of the remaining 22 patients, 2 patients expired in the follow up period. 20 patients follow-up data as available for analysis. Of these 7 were neonates, 8 infants and 5 patients were children older than one year.

Of the two patients who died, one was a neonates and the other was an infant. One died two months following surgery and had associated downs syndrome. This patients had undergone anorectalmyectomy one month after surgery due to constipation and distention. The second patient died 15 days after pullthrough surgery. Both the patients had ganglion cell in their proximal margin of resection. Both the deaths occurred outside the treating hospital and were being managed by local practitioner at the time of death.

Table 11: Early complication

	Perianal excoriation	Duration of excoriation	Increased frequency of stool	Duration of frequency
Neonates (n=7)	5(71%)	2.3 months	7(100%)	3.2 months
Infants (n=8)	5(62.5%)	2.1 months	5(62.5%)	2.5 months
Children (n=5)	2(40%)	1.5 months	3(60%)	2.2 months
Total (n=20)	12(60%)	1.96 months	15(75%)	2.63 months

Perianal excoriation was a problem in 60% of post operative patients. Excoriation was more common in neonates and resolved later than older children. Mean duration of excoriation in the study group was less than 2 months. Persistent excoriation was noted in two patients (one neonates and one infants) even after 20 months following surgery due to persistent soiling.

Increased frequency of stools (more than 5 to 7 per day) was noted in all neonates in the immediate postoperative period. Problem of increased frequency of stools after surgery was 2.6 months, prolonged increased frequency was noted in two patients (one infants and one child) up to 12 months following surgery.
One patient out of 30 operated upon had minor wound infected with partial wound dehiscence which was managed conservatively.

Table 12: frequency at last follow up

	1-2 stools/ day	2-4 stools / day	= 5 stools/ day
Neonates (n=7)	7(100%)	---	---
Infants (n=8)	4(50%)	2(25%)	2(25%)
Children (n=5)	3(60%)	1(20%)	1(20%)
Total (n=20)	14(70%)	3(15%)	3(15%)

All neonates and majority of the remaining patients have one to two stools per day at the time of last two follow up. 15% of the patients with stools of more than 5 per day are the lines with problems of incontinence and soiling.

Table 13: Late complications

	Stricture	Soiling	Enterocolitis	constipation	Death
Neonates (n=8)	2(25%)	---	---	---	1(16.6%)
Infants (n=9)	2(22.2%)	4(44.4%)	2(22.2%)	3(33.3%)	1(11.1%)
Children (n=5)	1(20%)	2(40%)	---	---	---
Total (n=22)	5(22.7%)	6(27.2%)	2(9%)	3(13.6%)	2(9%)

Stricture at the coloanal anastomotic site was noted in 5 patients (23%). Soiling was a problem in 6 patients (27%). Post-operative enterocolitis was noted in 2 Patients (9%). Occasional constipation was a complaints in 14% of the patients.

Table 14: additional surgical procedures

Procedure	Number
Anal bougeinage	6
Anal dilatation under anesthesia	2
Internal sphincterotomy	1
Anorectalmyectomy	1
Re-do surgery	1

Of five patients with anastomotic stricture two underwent dilation under general anesthesia and the remaining three patients were managed by regular anal bougienages. 4 out of these 5 patients are having normal bowel habits while one is incontinent with frequency soiling and episodes of enterocolitis.

The very first patients operated did not have a posterior myectomy done during surgery. Postoperatively the child had abdominal distension and required enema for evacuation, initially anal bougienage was attempted without symptomatic relief. Internal sphincterotomy was done one month after pull through procedure and anal dilatation was stopped. The child has normal bowel habits at present.

Anorectalmyectomy was done in one patient duo to persistent obstructive symptoms postoperatively who died 2 months after surgery. Single stage duhamel's procedure was done in one patients who had incomplete resection of the agaglionic segment of bowel during first pull through procedure.

Function result

Shankar et al in 2000[26] developed an analogue scoring system for patients of hisschsprung's disease which has been used to provide a functional outcome score in our patients.

Table 15: Functional scoring

Score	Neonates (n=7)	Infants (n=8)	Children (n=5)	Total (n=20)
1.normal bowel habits	6(85.71%)	4(50%)	2(40%)	12(60%)
2. soiling <1/week	1(14.28%)	0	2(40%)	3(15%)
3.soiling > 1/week	0	0	0	0
4. daily soiling or need for enema	0	3(37.5%)	1(20%)	4920%)
5.ACE, permanent stoma or major revision surgery		1(12.5%)	0	1(5%)

Normal bowel function was achieved in 85% of neonates and about 50% of older children. Daily soiling with need for enema was seen in 20% of the cases. One child underwent a revision surgery following incomplete resection of aganglionic segment.

According to this scoring system a satisfactory outcomes was defined as a score of 1 or 2 and poor outcome was defined as score of 3, 4 or 5.

Table 16: functional outcomes

Result	Neonates(n=7)	Infants(n=8)	Children(n=5)	Overall(n=20)
Satisfactory (1,2)	7(100%)	4(50%)	4(80%)	15(75%)
Poor (3,4,5)	0	4(50%)	1(14.3%)	5(25%)

75% of the study group have good functional outcome whereas remaining 25% had poor functional outcomes.

5out of 20 patients(20%) are older had 3 yrs of age, by which time continence is expected in children. Three patients (60%) are fully continent with normal bowel habits. One child is continent but complains of very occasional soiling. One child is having daily frequent soiling with no awareness of defecation.

IV. Discussion

Surgical treatment of hirschsprung's disease has changed significantly during the last decades. Multi stage surgery has progressed to open or laparoscopically assisted 1 stage repair. One stage totally transanal procedure is the latest evaluation in the management of hirschsprung's disease.

Primary endorectal pull through in the new born period was first described by so et al in 1980[18]. An 18 year follow up of these patients was reported recently and 81.5% were totally continent.

The rationale for primary surgery in the neonatal period has been the potential benefit of avoiding colostomy and establishment of colonic continuity early in life. This may enhance the chances of developing normal continence.

The incident of hirschsprung's disease among male patients compared to female in most literature is 80%. In our study males were slightly more 87% male preponderance in comparison.

The percentage of neonates in our study group was 33.3%. various centers in the world follow the principle of maintainingneonates on rectal washes to relive the function obstruction till such time that they have attained the age of 3 months before a repair is undertaken. In our study patients were taken up for pull through procedure as and when they presented to us after ensuring satisfactory decompression by means of rectal washes.

The mea age at which surgery was performed in neonates was 8.7 days. Infants were operated at the mean age of 5 months and in older children the mean age of surgery was 4.3 years.

All neonates were above 2.5 kg weight at the time of surgery. Larger et al [21]reported children weighing less than 4kgs were more prone o complications compared to children weighing greater then 4kgd. This

principle was followed by some and surgery was deferred till the child weighing 4kgs. This theory upon neonates with good results and no added complications. We operated upon the neonates and no additional complications were observed.

Babies of hirschsprung's disease are usually term deliveries as also seen in our study group.

As a historical finding delayed passage of meconium was very specific on neonatal age group. Distension was the commonest complaints across all age groups. In children above on year of age chronic constipation and chronic use of laxative was universal complaint.

Enterocolitis in our preoperative patients was seen only in 10% of cases which is low compared to western literature where figure of 19.5 %(waster and rintala) and 14.7% (Goa et al)[29] are quoted.

All neonates were symptomatic since birth while three-fourth of the infants and half of the children were symptomatic since birth. In children the onset of symptoms was delayed by 3 years (mean) range being 1-5 years, where as in infants symptomatology appeared at 3 months (mean) in whom delayed presentation was seen.

On clinical examination of patients in our study group, distension was seen in90% of the patients. The next commonest finding being visible or palpable loops (50%). The palpable or visible loops were more a feature in neonates and infants compared to children. This could be due to the thin and lax abdominal wall in neonates and infants compared to older children.

Similarly visible peristalsis was seen only in neonates (30%). Palpablefecaloma as one would expect impaction in long standing functional obstruction in hirschsprung's disease.

Explosive passage of stools on digital rectal examination is a finding typical to hirschsprung's disease. It was present in 50% of the neonate's and was not well elicited in patients who were older.

In our series 30% of the children and infants presented with pallor on clinical examination. After estimation of hemoglobin the incidence of anemia was higher that is 42% in older children and 30% in infants. Long standing obstruction in infants and children's results in nutritional deficiencies thereby leading to anemia.

One patient with associated down's syndrome was the only associated anomaly in our study sample. In a reported series by wester and rintala associated anomalies were seen in 10 to 40 patients. 8 patients had down's syndrome 3 of whom also had associated congenital cardiac defects (ASD, VSD). One patient had cartilage hypoplasia and one patient had central hypoventilation syndrome.

Contrast enema was diagnostic in all our patients with recto sigmoid disease being 63.33% in our study group. A comparative study between radiographic transition zone and level of aganglionosis conducted by Protocal et al, revealed that contrast enema showed a transition zone suggestive of Hirschusprung's disease in 67 of 75 patients the level of aganglionosis in 89% patients with rectosigmoid disease but only 31% in patients with long segment or total colonic HD may be encountered in 10% of patients with a rectosigmoid radiographic transition zone. With the advent of single stage procedure where contrast enema findings play a pivotal role it is vital to recognize this difference.

Most authors apart from contrast enema to know the level of aganglionosis also performed rectal biopsy to confirm the diagnosis of Hirschsprung's disease prior to pull-through procedure. Rectal biopsy was not done in our series as it is our belief that a well defined transition zone and a proximal dilated bowel on contrast enema has no other differential diagnosis apart from Hirschusprung's disease. Rectal biopsy was deliberately avoided as it would result in adhesions and subsequent difficulty in mucosal dissection during Transanal pull-through procedure. Some authors to circumvent this problem practiced full thickness rectal biopsy which was subjected of frozen section examination before starting pull-through surgery, but again frozen section is not 100%accurate.In our study no frozen section was used.

Mucosal dissection was generally easier in neonates compared to older children. Difficult mucosal dissection which had influenced the management was experienced in two of our patients. One patient had to be converted to Duhamel procedure and other resulted in incomplete resection of aganglionic segment. Submucosal dissection is difficult in older children because of thickness of mesentery, previous recurrent episode of enterocolitis, long standing dilated hypertrophied colon and previous rectal biopsy or anorectal myectomy.[27]

The length of intestine in our study was 12.3cms(mean), range being 6 to 20cms. The length of intestine resected was more in infants and children compared to that in neonates. The length resected in various other reported series is considerably more. Elhalaby and Elhalabary resected 15 to 45cms length of bowel in their series of 149 patients. 73% of their series had rectosigmoid disease and 9 patients had more proximal level of aganglionosis. Gao et al[29] in their series of 33 cases resected 29.5cms (mean) length of intestine range being 12.5 to 41cms. A. Haididi in his reported experience in 68 patients removed 25cms mean length of colonic segment (range 12 to 62cms). Hadidi removed any redundant colon proximal to transition zone. Cadaveric dissection showed that the lower one third of the descending colon could be dissected and pulled out of the anus because of its loose fixation to retroperitoneum (Teeratkul 2003).[27]

In our series 63.33% of the patients had rectosigmoidHirschsprung's disease and in remaining patients the transition zone was confined to rectum. Neonates and infants constituted two-thirds off our patient group and

redundant colon was not excised in our series. These could be factors responsible for resection of shorter segments compared to other studies.

There were no intraoperative complications in our 30 operated cases. Intra operative complications are rare in reported series.Hadidi[24] reported a causes of urethral injury. One author faced the problem of retraction of mesenteric vessel in two patients during ligation that required laparotomy to control bleeding twisting of bowel is a possibility at the time of pull-through.

The requirement of blood transfusion in our study was 53.33% of total patients. This was more compared to other reported series. Pre-operative poor nutritional status and preexisting anemia in 30% of infants and 42 % of children could be responsible. The general consensus is that mucosal dissection has to be carried out in the correct plane between mucosa and submucosa to minimize blood loss. Elhalaby et al in their study found blood loss to be greater in patients greater than one year of age compared with those less than one year of age (25% versus 14%). Similar findings were noted by Hadidi as was also seen in one study.

Feasibility of early postoperative feeding and short hospital staypostoperatively was comparable to various studies by other authors. Older children tolerated feeding earlier than the neonates and infants.

Early postoperative complication of perianal excoriation and increased frequency of stools was seen in 71% and 100% of the neonates in our study group respectively. Incidence of excoriation was inversely proportional to the age of the patient. Incidence of excoriation was lower in older patients. The overall incidence of excoriation in our series was 60%. In elhalaby's series of 149patients the incidence was 33%. In waster and rintala series 66% in neonatal age group and 36% beyond neonatal age group had excoriation. Langer et al reported an incidence of 47%. The increased incidence of excoriation in neonatal age group is a universal phenomenon noted by various authors as a result of sensitive skin in neonates.

Transient incontinence in immediate post operative period was seen in all neonates, 62.5% of infants and 60% of children. Overall incidence of transient incontinence was 75%. Rintala in his study of 26 patients noted transient incontinence in all patients Which resolved in few months. Two of his patients had increased frequently of stools in all his patients 4-6 weeks after surgery. In our study perianal excoriation lasted less than two months and two patients had persistent excoriation even after 20 months following surgery. Increased frequently of stools resolved after mean duration of 2.6 months following of more than 5 stools per day at the time of last follow up.

Minor wound infection was seen in one of our patient. Wound infection is rare after pull through procedure in most reported series. Peri-operative antibiotics and total gut irrigation in all our patients which prevents fecal contamination of pre operative site and decreases the bacterial load of gut are responsible for preventing would infection in our patients.

Complication like, anastomotic leakage, cuff abscess, peritonitis and prolapse of pulled through bowel as reported in other studies was not experienced in our study. Anastomotic leakage has been reported by Elhably, Langer and Hadidi. Hadidi had suggested anastomosis under tension and ischemia as probable causes for anastomotic leakage. All these patients mentioned in literature required diversion and subsequent repeat surgery. Redundant colon was through to be responsible for prolapse of pulled through bowel in early post operative period. Hadidi advocates excision of any redundant colon during pull-through procedure.Prolapse of pulled thorough colon can be treated by transanal excision of prolapsed colon and coloanal anastomosis. Though excision of redundant colon was not practiced by our surgeons, none of our patients developed prolapse of pulled through colon.

Regarding late complications, anastomotic stricture (25%), enterocolitis (9%), occasional constipation (14%), soiling (27%) and death (9%) was seen in our study group.

Incidence of anastomotic stricture was high in our series compared to other studies. Elhababy in his series reported stricture in 5% (7/149) patient requiring dilatation under general anesthesia and remaining % (13/149) who had stricture were managed by anal bougienage. In our series two out of five patients with anastomotic stricture required anal dilatation under anesthesia while the remaining three patients were managed with anal patients. One patient required 2attemts at stricturoplasty, another required stoma formation and repeated dilation, rest five patients responded to anal dilatation.

Postoperative enterocolitis was seen in two patients (9%). This is much less than that is experienced by other authors in western literature with reported incidence of as high as 32 to 42%. Elhalhaby, Langer[14], Rintala and Liu et al reported an incidence of 18%, 22%, 18%, and 24% respectively. Van leeuwen at al reported a relatively higher incidence of 56% of postoperative enterocolitis in his series. The wide variation could reflect the lack of standard defining criteria to label enterocolitis.

According to Elhalaby and Hadidi short muscle cuff, posterior myectomy and postoperative anal dilatation can help in reducing the incidence of post operativeenterocolitis. The two patients with enterocolitis in our study were managed by antibiotic for their treatment.

There were two moralities in our series which is comparable to other series. Elhalaby reported 3 deaths of 149 patients, De la Torre and Ortega Salgado reported 1 death of 10 patients treated with TEPT and 1 of 9 patients in Langer et al[21] series died.

Constipation requiring occasional laxatives or enema was encountered in 3 patients (13%). All these 3 patients had historically proven presence of ganglion cells in their pulled through colon. Leeuwen et al reported a very high rate of constipation, 35% (6 of 17) in his study.

One patient underwent single stage Duhamel procedure as re-do surgery as aganglionic segment of bowel was pulled through during initial TEPT. In two stage series by Langer[25] and Elhalabyinvolving 141 and 149 patients respectively re-do surgery was required in 2 patients in each of the series.

In Langer's series twisted pull-through and residual aganglionosis was cause for second surgery while in Elhalaby's series hypoganglionosis on permanent slides with severe constipation resulted in re-do surgery. One patient in our study group developed symptoms of obstruction in whom posterior splitting of muscular cuff was not done. This child underwent internal sphincterotomy and is presently having normal bowel function. One child underwent anorectalmyectomy for suspected sphincter achalasia. This child had associated Downs syndrome and expired 2 months post surgery due to unrelated cause.

Functional out come in our study group was satisfactory in 75% of the patients across all age groups. Satisfactory outcome was achieved in 100% of the neonates. Early outcome results were reported to be excellent in initial series but they concentrated more on disappearance of constipation rather than soiling or incontinence in patients after pull-thorough procedures. Shankar at al[26] reported early outcome result of 76% from data collected from Helsinki and Elhalaby in a multicenter study of 149 patients, but this was calculated in patients above the age of three years. 7 of his patients continue to have soiling and frequent accidents.

Langer et al[25] in his multicenter experience with 141 patients had reported 81% normal bowel function foe age. In a 18years follow up study of primary pull-through, in neonates by So et al total continence of 81% was reported.

In our study soiling (27%) and incontinence was a major complication compared to other studies. A lot of debate exists regarding the fact that over stretching of sphincter muscles during transanal pull-through procedures affect continence. Studies conducted by Leeuwen and others did not find any difference in anal manometric and rectoanal inhibitory reflex studies in post pull-through patients operated via a primal approach or incidence of constipation or enterocolitis and presence or absence of rectoanalinhibioty reflex. Though no follow up manometric study was done in our patient, all patients with incontinence or soiling had good anal tone on per rectal examination.

Authors are of the opinion that continence status improves with time and 10 years follow-up is required to correctly assess the functional status of operated patients.

V. Conclusion

A number of operative strategies have been described for Hirschsprung's disease. All are perceived to have relative merits and weakness, supported in some cases by medium and long term outcome data. Evaluation of different techniques is complex, and results of large single-surgeon series may reflect the merits of the surgeon rather than the operation. Randomized controlled trails are not feasible, and where an institute has adopted a new technique, only historical comparison is possible. In contrast to functional outcome is complicated. Lack of consensus regarding socially acceptable norms, relative insensitivity of questionnaire tools, and lack of generally accepted functional continence score all contribute to this. The inevitable delay between treatment and assessment of continence at age of 3 to 4 years prolongs the process of comparison.

Initial results reported in literature after single stage transanalendorectalpullthrough were very encouraging and result were comparable to the time tested staged procedures. The results were based on small sample size and relief of obstruction was considered as satisfactory outcome. Recent reports of medium term follow up based on larger group of operated patients place good to satisfactory outcome at 75 to 80%. The problems of soiling, incontinence and in some cases constipation in significant number of patients with this new technique has been highlighted by few authors. The general opinion is that a careful long term followup is required to determine whether patients clearly are benefiting from this techniques. As seen in our study after mean follow up of about 18 months that problem of soiling and incontinence are real and cannot be over looked.

Due to its numerous advantages like avoiding colostomy, no peritoneal breach, preservation of pelvic innervations, avoiding injury to pelvic organs, excellent cosmesis, early feeding, short hospital stay, cost effective, less separation from parents, less operative time, lesser blood loss and less pain it is very attractive option. In contrast to transabdominal approach the risk of developing adhesive obstruction in transanal approach is minimal. The reported incidence of adhesive obstruction after open pull through for HD is 2% to 20% which is quite significant. In contrary to open procedures no urinary complications has been reportedfollowing TEPT procedure as dissection always confined within the rectal wall thereby avoiding injury to pelvic innervations.

The argument put forth in favor of TEPT is that is not a new surgical but a new approach, so if transabdominal pull-through has stood the test of time for all practical purpose the results of proving that over stretching of anal sphincters at the time of surgery in perineal approach does not compromise sphincter integrity also goes in favor of TEPT.

As seen in our study and also by other authors the results of those procedure are better in neonatal period compared to other age groups. Good outcomes could be achieved in all neonates in our study. Moreover younger patients show steady improvement in their continence status over passage of time compared to older children. Taking learning curve into consideration for all new techniques better results are expected in future as more experience is gained.

Thus his new technique with its all advantages has a role in surgical management of Hirschsprung's disease especially in neonates where a definitie transition zone at the recotosigmoid level could be identified.

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