

**SINGLE STAGE TRANSANAL ENDORECTAL  
PULL-THROUGH (T.E.P.T) FOR  
HIRSCHSPRUNG'S DISEASE**



*Dissertation Submitted to*  
**Coimbatore Medical College & Hospital,  
Coimbatore**

*For*  
**M.Ch. - Paediatric Surgery  
Branch – V**



**The Tamil Nadu  
Dr. M. G. R. Medical University  
Chennai**

**AUGUST – 2009**

## **CERTIFICATE**

This is to certify that this dissertation titled '**SINGLE STAGE TRANSANAL ENDORECTAL PULL-THROUGH (T.E.P.T) FOR HIRSCHSPRUNG'S DISEASE**' is a bonafide work of **Dr.L.SENTHIL KUMAR**, submitted for the qualifying examination in M.Ch., Paediatric Surgery, to be held in August 2009 by the **Dr. M.G.R. MEDICAL UNIVERSITY.**

Signature of the H.O.D

Signature of the Dean

## **ACKNOWLEDGEMENT**

I wish to record my sincere thanks to our **PROF.V. KUMARAN MS.,M.Ch., DEAN I/C Coimbatore medical college hospital, Coimbatore** for permitting me to work for this Dissertation and to avail all the facilities in this Institution.

I am deeply indebted to our **PROF. V. KUMARAN, M.S., M.Ch.**, Professor and Head of the Department of Paediatric Surgery, Coimbatore Medical College Hospital, Coimbatore, but for whose guidance, this study would not have come through. It has been a great privilege to work under him and especially on this topic.

I express my profound gratitude to **Dr. G. RAJAMANI, M.S., M.Ch., Dr. S. KANNAN, M.S., M.Ch., Dr. N. VENKATESA MOHAN, M.S., D.N.B, Dr. R. RENGARAJAN M.S., M.Ch., Dr. M. NATARAJ, M.S., M.Ch.& Dr.V.MUTHULINGAM M.S.,M.Ch.**, for their generous help, advice and suggestions in various stages of this study.

This study would not have seen the light of the day, had not our patients showed the kind co-operation they extended. I sincerely thank them.

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## INTRODUCTION

Hirschsprung's Disease (HD), named after Harold Hirschsprung, who presented the classic description in 1886,<sup>1</sup> 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.<sup>5,6</sup> It usually presents 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 cholinergic and adrenergic excitatory innervations 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 acetylcholinesterase 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.<sup>7,8,9</sup> First successful surgical technique was described by Swenson and Bill. A retrorectal approach described by Bernard Duhamel in 1956<sup>14</sup> and endorectal pull through technique described by Franco Soave in 1960<sup>15</sup> and subsequently modified by Boley<sup>16, 17</sup> 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. These three stage

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 complications lead 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 in 1995.

Primary transanal endorectal pull-through was first described in 1998.<sup>20</sup> 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 adhesive 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 procedures 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 procedures have been adapted to minimally invasive approach. All use at least three abdominal port sites with the associated risks of bleeding and thermal or harmonic injury to other pelvic structures. Laparoscopic surgery requires adequate expertise, experience and assistance. Single stage transanal endorectal pull-through provides the same advantages as laparoscopic surgery with additional advantage of eliminating risk of intraabdominal pelvic dissection and adhesion formation, no scars and less pain.

Various studies over last few years have reported the results of transanal endorectal pull-through as good as, if not better than staged procedures.<sup>32,33,34</sup> Single stage transanal pull-through was started in our institute in year 2002. 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 transanal endorectal pull-through for rectosigmoid Hirschsprung's Disease.

## **AIMS OF THE STUDY**

1. To evaluate the functional results in children with Hirschsprung's Disease who had undergone Single Stage Transanal Endorectal Pull through.
2. To evaluate the stooling pattern post operatively in these patients.
3. To study the immediate and late complications.
4. To assess patient and parental satisfaction.

## **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 Patients 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 Patients 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 6 yrs of age.

This 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 and hospital, Coimbatore from Jan 2006 to Jan 2009 .

All patients with history suggestive 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 segment, a funnel shaped transition zone and proximal dilated 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 transanal endorectal pull-through procedure (TEPT). More proximally located transition zone, long segment Hirschsprung's disease and total colonic aganglionosis were managed by staged procedure.

Patients 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 Transanal Endorectal 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

1Preoperative clinical data (presenting signs and symptoms) and diagnostic studies.

2Operative details including type of pull through, length of bowel resected, intraoperative complications and blood transfusion requirement.

3Early postoperative course like degree of pain, timing of starting oral feeds

4Early and late complications

5Additional surgical procedures, if any.

6Functional outcome as judged by bowel habits and anorectal continence

7Histopathological findings.

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

#### **Pre-operative preparation:**

Once bowel decompression was ensured by frequent rectal irrigation, mainly in infants and older children, patients were planned for Single Stage Transanal Endorectal Pull through. The day prior to surgery every patient received total gut irrigation. Through a naso-gastric tube normal saline at a rate of 25ml/kg/hr was infused along with intermittent rectal washes till the effluent per anum was clear and free of fecal residue. This usually took 4-6 hours.

## **Surgical Technique:**

After induction of anaesthesia, the patient was placed in lithotomy position with pelvis elevated at the end of the operating table. Intravenous antibiotics, often a third generation cephalosporin was given at induction and bladder was catheterized by an indwelling catheter. Naso-gastric tube inserted for total gut irrigation was maintained.

Mini Laprotomy done in Left Iliac fossa, Transition zone identified, Marker stitch taken about 3-5cm above Transition zone at antimesentric border. In some cases we done Laproscopy 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.



Figure 1. Anal canal exposed by stay sutures

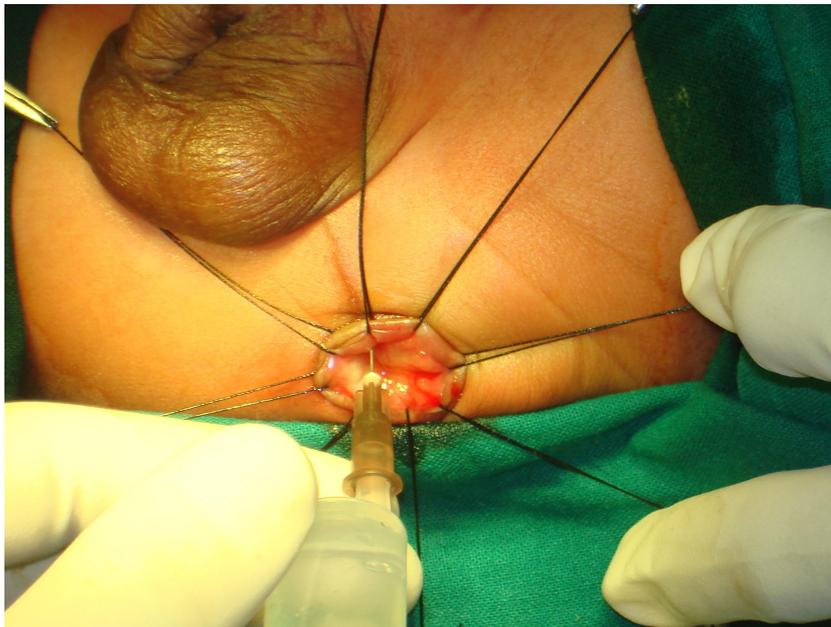


Figure 2. Submucosal saline injection

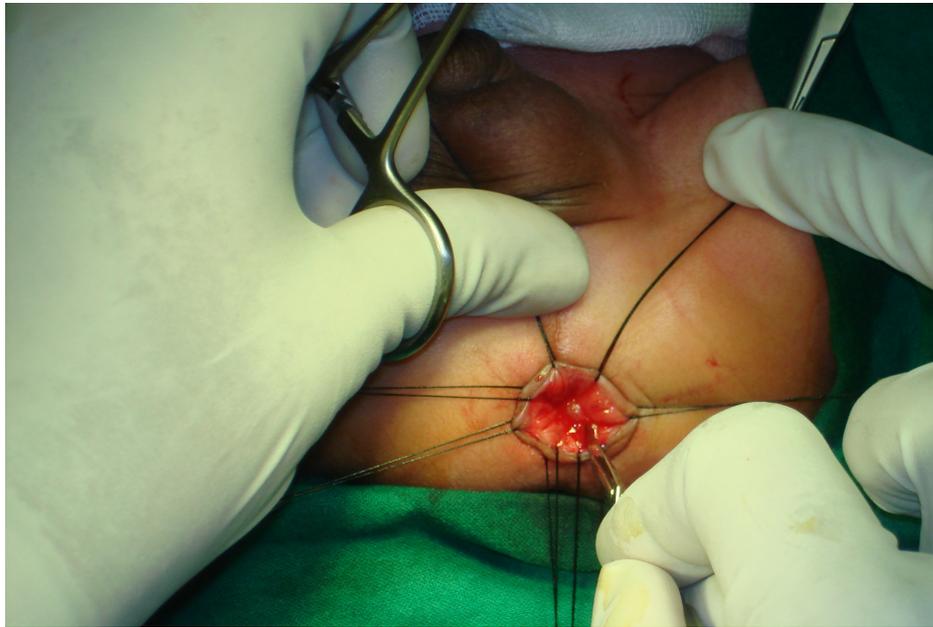


Figure 3. Incision of rectal mucosa.

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 extension of mucosal dissection until the level proximal to peritoneal reflection.



Figure 4. Mucosal tube dissection.

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

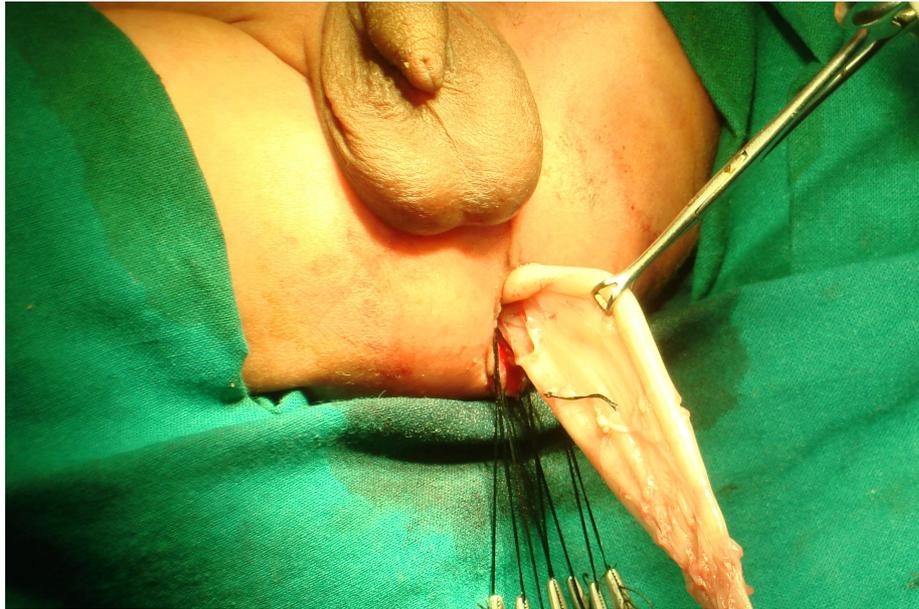


Figure 5. Mobilization of full thickness colon & Identification of Marker stitch.

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



Figure 6. Muscle cuff incision posteriorly.

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 polygalactin 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 started 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 was 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 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 visit was usually 2 weeks after surgery. Later, regular follow up 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 dilatation was not carried out for initial few patients. Due to occurrence of anastomotic strictures in some of the initial patients, We resorted to routine anal dilatation starting one month after surgery.

No routine follow up investigations were done in our study.

## RESULTS

A total of 30 patients were included in this study from January 2006 to January 2009. Of these patients 26 were male patients (86.6%) and 4 were female patients (13.3%).

**Table 1: Age profile of the patients**

<b>Patient group</b>	<b>Number</b>	<b>Percentage</b>
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**

<b>Patient group</b>	<b>Mean Age (Range)</b>	<b>Mean weight (Range)</b>
Neonates	8.7 days (3-20)	2.87 Kg (2.5-3.3)
Infants	5.11 months (2-12)	5.62 Kg (2.7-9.5)
Children	4.35 years (2.5-6)	12.62 Kg (9.2-16.0)

All the patients were term deliveries with no specific perinatal complications.

All neonates were of good weight, mean weight being 2.87kg (2.5-3.3)

**Table 3: Presenting symptoms**

<b>Symptoms</b>	<b>Delayed passage of meconium</b>	<b>Distension</b>	<b>Vomiting</b>	<b>Chronic constipation</b>	<b>Chronic laxative Use</b>	<b>Enterocolitis</b>
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(100%)	1(14.28%)
Total (n=30)	25(83.3%)	29(96.6%)	13(43.3%)	21(69%)	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**

<b>Group</b>	<b>Since birth</b>	<b>Delayed</b>
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 (2-5months) in whom onset was delayed.

**Table 5: Clinical examination findings**

	<b>Distension of abdomen</b>	<b>Visible or Palpable Bowel loops</b>	<b>Visible Peristalsis</b>	<b>Palpable fecalomas</b>	<b>Explosive passage of stools on per rectal examination</b>	<b>Pallor</b>
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%)	10(33.3%)	7 (23.3%)

Abdominal distension was the commonest clinical finding in all age groups. Palpable or visible bowel loops was more commonly a feature in younger patients, where as 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 Down's syndrome apart from which no other associated anomalies were present in our study group.

### **Investigative results**

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

**Table 6: Contrast enema findings**

	<b>Rectum</b>	<b>Rectosigmoid</b>	<b>Sigmoid</b>
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 rectosigmoid junction, while transition zone confined to rectum was seen 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**

	<b>Length of resected bowel (in cms)</b>
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**

	<b>Number of patients who received transfusion</b>
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 these 3 patients one patient had continuous obstructive symptoms in the postoperative period. Repeat contrast enema of this child revealed persistent transition zone and this child further underwent a one stage Duhamel's procedure. The 2<sup>nd</sup> patient in this category is having normal bowel functions at 9 months of follow up. The 3<sup>rd</sup> patient has been lost to follow up.

In five patients 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, patients 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 of starting post operative oral feeds**

	Mean day of starting oral feeds (range)
Neonates (n=10)	2.7 days (2-4 days)
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**

	<b>Total hospital stay</b>	<b>Post operative hospital Stay</b>
Neonates (n=10)	12.4 days(9-17days)	5.1 days (4-7 days)
Infants (n=13)	9.23 days(5-23days)	4.1 days (3-6 days)
Children (n=7)	11.7 days(5-26days)	4.7 days (3-7 days)
Total (n=30)	10.8 days(5-26days)	4.5 days (3-7days)

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

### **Follow up period**

Mean follow up period is 17.3 months range being 5 months to 38 months. Of the total 30 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 was 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 neonate and the other was an infant. One died two months following surgery and had associated Down's syndrome. This patient had undergone anorectal myectomy one month after surgery due to constipation and distension. 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.

**Table11: Early complications**

	<b>Perianal excoriation</b>	<b>Duration of excoriation</b>	<b>Increased frequency of stool</b>	<b>Duration of frequency</b>
Neonates (n=7)	5 (71%)	2.3 months	7(100%)	3.2months
Infants (n=8)	5 (62.5%)	2.1 months	5(62.5%)	2.5months
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 neonate and one infant) 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 stool was less in older children. Mean duration of frequent stools after surgery was 2.6 months. Prolonged increased frequency was noted in two patients (one infant and one child) up to 12 months following surgery.

One patient out of 30 operated upon had minor wound infection with partial wound dehiscence which was managed conservatively.

**Table 12: Frequency at last follow up**

	<b>1-2 stools / day</b>	<b>2-4 stools /day</b>	<b>≥ 5 stools /day</b>
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 follow up. 15% of the patients with stools of more than 5 per day are the ones with problems of incontinence and soiling.

**Table 13: Late complications**

	<b>Stricture</b>	<b>Soiling</b>	<b>Enterocolitis</b>	<b>Constipation</b>	<b>Death</b>
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 complaint in 14% of the patients.

**Table 14: Additional surgical procedures**

<b>Procedure</b>	<b>Number</b>
Anal bougeinage	6
Anal dilatation under anaesthesia	2
Internal sphincterotomy	1
Anorectal myectomy	1
Re-do surgery	1

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

The very first patient 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.

Anorectal myectomy was done in one patient due to persistent obstructive symptoms postoperatively who died 2 months after surgery. Single stage Duhamel's procedure was done in one patient who had incomplete resection of the aganglionic segment of bowel during first pull-through procedure.

**Functional results:**

Shankar et al in 2000 <sup>26</sup> developed an analogue scoring system for patients of Hirschsprung's disease which has been used to provide a functional outcome score in our patients.

**Table 15: Functional scoring**

<b>Score</b>	<b>Neonates (n=7)</b>	<b>Infants (n=8)</b>	<b>Children (n=5)</b>	<b>Total (n=20)</b>
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/wk	0	0	0	0
4. Daily soiling or need for enema	0	3(37.5%)	1(20%)	4(20%)
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 outcome was defined as a score of 1 or 2 and poor outcome was defined as a score of 3,4 or 5.

**Table16: Functional outcome**

<b>Result</b>	<b>Neonates(n=7)</b>	<b>Infants(n=8)</b>	<b>Children(n=5)</b>	<b>Overall (n=20)</b>
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 outcome.

5 out of 20 patients (25%) are older than 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.

## REVIEW OF LITERATURE

Harold Hirschsprung presented the classic description of the disease entity in 1886.<sup>1</sup> Description of children with megacolon dates back to 17<sup>th</sup> century, Frederick Ruysch 1691.<sup>4</sup> However the appreciation of the disease did not occur till Hirschsprung's report.

The understanding of the pathogenesis of the disease took several more decades. Three basic theories were put forward. The malfunction theory, the obstructive theory and finally the spastic theory, put forward by Fenwick in 1900, who proposed for the first time that distal colon was spastically contracted to cause functional obstruction.<sup>2</sup> The absence of ganglion cells in distal colon of children with megacolon was first noted by Tittel in 1901.<sup>4</sup> In 1946, Ehrenpreis was the first to appreciate that the colon became secondarily dilated due to distal obstruction.<sup>3</sup> Over the decades as the understanding of the pathophysiology of Hirschsprung's disease improved, a more rational approach to its diagnosis and treatment developed.

Surgical technique has undergone a sea of changes over the last century initially through trial and error and subsequently for better patient care and to reduce complications of surgery. The

basic principle though remained the same that is to resect the aganglionic segment and maintain continuity with peristaltic bowel.

The first surgical treatment offered was a diverting colostomy, but symptoms returned after closure of the ostomy. Attempts at bypass or removal of the redundant colon were uniformly unsuccessful. Ladd and Gross reported improvement in symptoms after lumbar sympathectomy to negate the sympathetic input to the rectum.<sup>7,8,9</sup> First successful surgical approach was by Swenson and Bill in 1948.<sup>10</sup> A circumferential anastomosis was performed at the level of internal sphincter after resection. High incidence of incontinence and stricture lead to modification by Swenson, which involved a more oblique anastomosis with sparing of internal sphincter. A low anterior resection of the rectosigmoid in the State procedure<sup>11</sup> that did not resect sufficient length of aganglionic intestine has been abandoned. Subsequent modification of this technique of low anterior resection by Rehbein in 1953<sup>12,13</sup> who resected the bowel well beyond the peritoneal reflection distally, is still successfully used. Bernard Duhamel described his operation for Hirschsprung's Disease in 1956.<sup>14</sup> A retrorectal approach was used which involved minimal pelvic dissection thereby preserving sensory innervations to rectum.

Franco Soave introduced the endorectal pull-through technique in 1960.<sup>15</sup> This consists of removal of mucosa and submucosa of the rectum and ganglionated intestine is pulled through the aganglionic muscular cuff. Boley further modified this technique by performing a formal anastomosis at the anus.<sup>16,17</sup>

Various modification of these three techniques namely Duhamel's, Soave's and Swenson's gained worldwide popularity and became the procedure of choice for Hirschsprung's Disease in most of the centers.

The initial procedures were three-staged which was subsequently converted to a two-staged one by foregoing a protective colostomy for the definitive surgery. The main disadvantages of staged procedures are multiple hospitalization, multiple exposure to anesthesia and foremost being extended period of colostomy. Difficulties in managing a patient on colostomy for almost a year with its associated complications and quest for a stoma free treatment for Hirschsprung's Disease lead to single stage procedures.

In 1980, So et al reported about 20 newborns who were treated with endorectal pull-through and no previous colostomy.<sup>18</sup> With the advent of minimally invasive technique in pediatric

surgery the treatment of H.D could not have stayed unaffected. Laparoscopic mobilization of colon and pull-through was forwarded by Georgeson et al in 1995.<sup>19</sup>

De la Torre-Mondragon and Ortega-Salgado<sup>20</sup> in 1998 described mucosectomy, colectomy, and pull-through using the transanal approach without the need for laparotomy or laparoscopy in the management of rectosigmoid H.D. This was followed by experiences from other centers, Langer et al<sup>21</sup> and Albanese et al,<sup>22</sup> in 1999. Over the next five years this technique got recognition and many centers adopted this method to treat rectosigmoid Hirschsprung's Disease as a first line. Fabio et al in 2003 described a modified technique of primary transanal rectosigmoidectomy, using a Swenson like procedure to perform the anastomosis between colon and the rectum.<sup>23</sup>

One of the essential limitations of an entirely TEPT is the proximal extension of the ganglionic segment beyond the sigmoid colon. Cadaveric dissection showed that lower one third of the descending colon could be dissected and pulled out of the anus because of its loose fixation to the retroperitoneum.<sup>27</sup>

Because of the possibility of conversion to laparotomy when a histologically proven transition zone cannot be reached from below,<sup>42</sup> the supine position is recommended rather than the prone position, which is preferred by some surgeons with the assumption of better control of mesenteric vessels.<sup>20</sup> Conversion to laparotomy should not be considered a failure of TEPT technique, and the basic principles of treatment of HD, which entails resection of the aganglionic segment and bringing a normally innervated bowel down to anus, should never be violated at the expense of avoiding laparotomy.

The remaining seromuscular cuff after TEPT has been accused for the development of postoperative obstructive symptoms, constipation, and enterocolitis.<sup>29</sup> The cuff may constrict around the pulled through bowel or even roll down and cause anorectal narrowing. Langer et al<sup>21</sup> and Albanese et al<sup>22</sup> left a long cuff, but cut it posteriorly. Elhalaby<sup>39</sup> prefers to continue mucosectomy for a relatively long length to be sure that intraabdominal part of rectum is reached, to avoid injury to ureter and or vas deferens, which are at risk if full thickness dissection is started below peritoneal reflection. The cuff is then inverted outside the anus and shortened to the exactly needed length

before returning it back to its normal position. Rintala et al<sup>31</sup> leaves a very short cuff of 3 to 4cms length without splitting the cuff posteriorly.

One of the crucial critiques for the TEPT approach is the significant stretching of the anal sphincters during mucosectomy with its potential impact on postoperative continence status particularly in older children with marked hypertrophy and dilatation of the colon.<sup>34,42</sup> Current studies show that transient soiling and increased frequency of bowel motions does occur in significant number of patients probably because of stretch effect. However this proved to be a transient effect and bowel movements became normal in the majority of cases within a period from 2 weeks to 3 months. Leeuwen et al<sup>34</sup> reported that anorectal manometric studies were similar in both patients who had TEPT or conventional endorectal pull-through. Elhalaby<sup>39</sup> found postoperative EMG mapping of anal sphincters to be within normal range in those patients who had frequent soiling and incontinence.

Another critical issue is related to the relatively distal level of rectoanal anastomosis. A low anastomosis at or distal to the dentate line may damage the delicate nerve endings that play a part in anorectal continence.<sup>42</sup> Patients with lower anastomosis at

or distal to the dentate line were associated with higher frequency of transient soiling for more prolonged periods than in those with more proximal anastomosis.<sup>39</sup> Anorectal continence is expected to be favorable in younger patients who continue to show steady improvement of their continence status over period of time.

Regarding post operative anal dilatation Langer et al<sup>21</sup> are of the opinion that anal dilatation or bouginage should be reserved for cases with existing or potential risk of stricture formation. Elhalaby et al<sup>39</sup> believe that postoperative routine anorectal bouginage is an effective tool to prevent the occurrence of anal stricture and to decrease both the frequency as well as the severity of enterocolitis particularly in neonates and young infants. Rintala et al<sup>41</sup> are of similar opinion and anal dilatations were required more frequently in neonates in their study. The author recommends careful early followup of neonatal patients and daily anal dilatation in patients with an anus tighter than Hegar size 12 two weeks after surgery.

Postoperative enterocolitis after TEPT has been variously reported from 0 to 56% in literature. This wide variation is due to lack of fixed criteria for diagnosing enterocolitis. Elhalaby et al<sup>37</sup> attempted to stratify the severity of enterocolitis into three grades.

Grade 1- mild explosive diarrhea, mild to moderate abdominal distension, no significant systemic manifestation.

Grade 2- moderately explosive diarrhea, moderate to severe abdominal distension associated with mild to moderate systemic manifestations (i.e. fever and tachycardia)

Grade 3- explosive diarrhea, marked abdominal distension, shock or impending shock

Patients with grade 1 enterocolitis can be treated successfully as out patients whereas those with grade 2 or 3 require hospitalization and colonic decompression along with intravenous antibiotics.

Teitelbaum et al<sup>43</sup> provided a histopathological grading of enterocolitis which is as follows:

- Grade 0 – normal mucosa
- Grade 1 – crypt dilatation or crypt with retained mucin
- Grade 2 – two or fewer crypt abscesses per high power field
- Grade 3 – multiple crypt abscesses
- Grade 4 – intraluminal fibrinopurulent debris or ulceration of the mucosal epithelium
- Grade 5 – transmural necrosis or perforation

It has been suggested that postoperative enterocolitis is the most reliable indication of the successful or unsuccessful relief of the low intestinal obstruction in Hirschsprung's disease.

Authors have described use of laparoscopy and laparotomy as an adjunct where ganglionic segments could not be reached transanally. Proctor et al<sup>36</sup> in their study found that long segment and total colonic Hirschsprung's disease may be encountered in upto 10% of patients with a rectosigmoid transition zone on contrast enema. Their recommendation is that, a biopsy confirmation of transition zone, via laparoscopy or mini laparotomy should be done before beginning rectal dissection. There is a debate whether routine laparoscopic visualization of transition zone is required for transanal procedures. Laparoscopy offers several benefits, including 1. determination of the level of transition zone before committing one self to perineal dissection, 2. visualization of pulled through bowel to ensure that there is no twisting or bleeding, 3. mobilization of splenic flexure if there is a more proximal transition zone. Disadvantages of laparoscopy are increased operating time, increased cost, need for expertise and potential risk of developing adhesive obstruction. Langer et al<sup>34</sup> in their study concluded that routine laparoscopic visualization or

mini-laparotomy is not necessary in patients planned for one stage pull through.

The proponents and opponents of laparoscopic- assisted approach have their own arguments. One compromise between the two is what is followed by Jean-Martin LaBerge of Montreal. He puts the scope in, does the biopsies, and then does the entire surgery transanally in all cases. This avoids laparoscopic pelvic dissection which carries certain risks and does not require expertise of laparoscopic pelvic dissection in a small child.

Apart from the obvious advantages of it being a single stage procedure, experience from other centers have shown TEPT to take less op-time, less bleeding, shorter hospital stay, less postoperative pain, early oral feeding, less separation from the parents and more cost effective. The surgery being totally transanal it gave the best cosmetic result with no abdominal scar. Peritoneum was not violated, obviating the chances of post-operative adhesive obstruction.<sup>14,21,24,25,28,34,35</sup> Dissection being transanal pelvic innervations are preserved and injury to other pelvic organs is avoided. In most reported series children attained a stool frequency of three per day by three to six months postoperatively. Complications like anastomotic leaks, stricture

formation, constipation and incontinence were either very less or absent.<sup>20,21,26,27,28,29,30,31,35</sup> Comparison of functional results between transanal primary pull-through and conventional trans abdominal approaches were found to be similar.<sup>32,33,34</sup> Though most series reported excellent early results with few complications, long term follow up is required to assess the exact bowel and sexual function.

## 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 evolution in the management of Hirschsprung's disease.

Primary endorectal pull through in the newborn period was first described by So et al in 1980.<sup>18</sup> A 18 year follow up of these patients was reported recently and 81.5 % were totally continent.<sup>38</sup>

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 incidence of Hirschsprung's disease among male patients compared to females 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 maintaining

neonates on rectal washes to relieve the functional obstruction till such time that they have attained the age of 3 months before a repair is undertaken<sup>39</sup>. 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 mean 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. Langer et al (1999)<sup>21</sup> reported children weighing less than 4 kgs were more prone to complications compared to children weighing greater than 4kgs. This principle was followed by some and surgery was deferred till the child weighed 4kgs. This theory lost its popularity as more and more authors world wide operated upon neonates with good results and no added complications. We operated upon ten 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 in neonatal age group. Distension was the commonest complaint across all age groups. In children above one year of age chronic constipation and chronic use of laxative was a universal complaint.

Enterocolitis in our preoperative patients was seen only in 10% of cases which is low compared to western literature where figures of 19.5% (Wester and Rintala )<sup>41</sup> and 14.7% (Gao et al)<sup>29</sup> 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 in 90% 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%). Palpable fecalomas as one would expect was a feature of older children (42%) due to the faecal 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 neonates 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 anaemia was higher that is 42% in older children and 30% in infants. Long standing obstruction in infants and children results in nutritional deficiencies thereby leading to anaemia.

One patient with associated Down's syndrome was the only associated anomaly in our study sample. In a reported series by Wester and Rintala<sup>41</sup> associated anomalies were seen in 10 of 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 Proctor et al,<sup>36</sup> revealed that contrast enema showed a transition zone suggestive of Hirschsprung's disease in 67 of 75 patients (89%). Contrast enema correctly predicted the level of aganglionosis in 89% patients with rectosigmoid disease but only 31% in patients with long segment or total colonic disease. According to their findings 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 procedures 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 zones, that is, a narrow distal segment, a funnel shaped transition zone and a proximal dilated bowel on contrast enema has no other differential diagnosis apart from Hirschsprung'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<sup>27</sup> to circumvent this problem practiced full thickness rectal biopsy which was subjected to 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.<sup>39,27</sup>

The length of intestine resected 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 Elbarbary<sup>39</sup> 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<sup>29</sup> in their series of 33 cases resected 29.5cms (mean) length of intestine range being 12.5 to 41cms. A. Haididi<sup>35</sup> in his reported experience in 68 patients removed 25cms mean length of colonic segment (range 12 to 62cms). Hadidi<sup>35</sup> 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)<sup>27</sup>.

In our series 63.33% of the patients had rectosigmoid Hirschsprung's disease and in remaining patients the transition zone was confined to rectum. Neonates and infants constituted two-thirds of 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<sup>24</sup> reported a case of urethral injury. One author faced the problem of retraction of mesenteric vessel in two patients during ligation that required laparotomy to control bleeding<sup>39</sup>. Twisting of bowel is a possibility at the time of pull-through.<sup>34</sup>

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<sup>39</sup> 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<sup>35</sup> as was also seen in our study.

Feasibility of early postoperative feeding and short hospital stay postoperatively was comparable to various studies by other authors<sup>39,41</sup>. Older children tolerated feeding earlier than the neonates and infants.

Early post operative 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<sup>39</sup> of 149

patients the incidence was 33%. In Wester and Rintala series<sup>41</sup>, 66% in neonatal age group and 36% beyond neonatal age group had excoriation. Langer et al<sup>34</sup> 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<sup>31</sup> in his study of 26 patients noted transient incontinence in all patients which resolved in few months. Two of his patients had increased frequency of stools beyond six months. Similarly Hadidi<sup>24</sup> noted increased frequency 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 frequency of stools resolved after mean duration of 2.6 months following surgery in our study. Only 3 patients had stool frequency 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 the operative site and decreases the bacterial load of gut are responsible for preventing wound infection in our patients.

Complications like, anastomotic leakage, cuff abscess, peritonitis and prolapse of pulled through bowel as reported in other studies<sup>39,41</sup> was not experienced in our study. Anastomotic leakage has been reported by Elhalaby<sup>39</sup>, Langer<sup>34</sup> and Hadidi<sup>35</sup>. 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 thought to be responsible for prolapse of pulled through bowel in early post operative period. Hadidi<sup>35</sup> advocates excision of any redundant colon during pull-through procedure. Prolapse of pulled through colon can be treated by transanal excision of prolapsed colon and coloanal anastomosis<sup>39</sup>. 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. Elhalaby<sup>39</sup> in his series reported stricture in 5% (7/149) patient requiring dilatation under general anesthesia and remaining 9% (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 bougienage. Minford et al<sup>40</sup> reported stricture in 7 of 37 (19%) of patients. One patient required 2 attempts 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%. Elhalaby<sup>39</sup>, Langer<sup>14</sup>, Rintala<sup>41</sup> and Liu et al reported an incidence of 18%, 22%, 18% and 24% respectively. Van Leeuwen et al<sup>33</sup> 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<sup>39</sup> and Hadidi<sup>35</sup> short muscle cuff, posterior myectomy and postoperative anal dilatation can help in reducing the incidence of post operative enterocolitis. The two patients with enterocolitis in our study were managed by antibiotics and anal dilatation and did not require hospitalization for their treatment.

There were two mortalities in our series which is comparable to other series. Elhalaby<sup>39</sup> reported 3 deaths of 149 patients, De la Torre and Ortega Salgado<sup>33</sup> reported 1 death of 10 patients treated with TEPT and 1 of 9 patients in Langer et al<sup>21</sup> series died.

Constipation requiring occasional laxatives or enema was encountered in 3 patients (13%). All these 3 patients had histologically proven presence of ganglion cells in their pulled through colon. Leeuwen et al<sup>33</sup> 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 large series by Langer<sup>25</sup> and Elhalaby<sup>39</sup> involving 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 anorectal myectomy 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-through procedures. Shankar et al<sup>26</sup> reported early outcome result of 76% from data collected from Helsinki and Liverpool hospitals. 83% complete continence was reported by Elhalaby<sup>39</sup> 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<sup>25</sup> in his multicenter experience with 141 patients had reported 81% normal bowel function for age. In a 18 years follow up study of primary pull-through in neonates by So et al<sup>18</sup> 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<sup>33</sup> and others did not find any difference in anal manometric and rectoanal inhibitory reflex studies in post pull-through patients operated via a perineal approach or transabdominal approach They found no correlation between the incidence of constipation or enterocolitis and presence or absence of rectoanal inhibitory 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.

## **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 trials are not feasible, and, where an institute has adopted a new technique, only historical comparison is possible. In contrast to establishing the safety of a new procedure, assessment of 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 Transanal Endorectal Pullthrough were very encouraging and results 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 patient places 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 follow up is required to determine whether patients clearly are benefiting from this technique. As seen in our study after mean follow up of about 18 months that problems 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 a 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 reported

following TEPT procedure as dissection is always confined within the rectal wall thereby avoiding injury to pelvic innervations.

The argument put forth in favour of TEPT is that it is not a new surgery but a new approach, so if transabdominal pull-through has stood the test of time for all practical purpose the results of TEPT should be no different. The fact that manometric studies 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 this procedure are better in neonatal period compared to other age groups. Good outcome 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 this new technique with all its advantages has a role in surgical management of Hirschsprung's disease especially in neonates where a definite transition zone at the rectosigmoid level could be identified.

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## Appendix I

### CONSENT FORM

Name:

IP No.

I hereby give my consent for my child to be part of the study

entitled "Single Stage Transanal Endorectal Pull-Through for Hirschsprung's Disease " .

I have been fully explained the procedure involved and I give my full and unreserved consent for participation in this study.

Witness:

Signature of parent /guardian

Date:

Place:

**PROFORMA**

**Appendix II**

Name : Age/Sex : IP.No:

Address : Tel.no :

Wt at presentation

Date of admission : Date of surgery : Date of discharge:

**Antenatal History :**

**Presenting Complaints:**

Delayed passage of meconium: Distension:

Vomiting: Constipation:

Duration of symptoms: Enterocolitis:

Use of laxatives:

**Clinical Examination:**

**INVESTIGATIONS :**

Hb/TLC : Urea/Creat: Na/K: Others:

**Radiology :**

Plain X-ray :

Contrast Enema :

**Rectal Biopsy:**

**Associated anomaly:**

**Diagnosis / Type:**

**Surgery :**

Findings:

Length resected:

Post myectomy

Operative time:

Blood loss:

Blood Transfusion:

Intra-operative Complication:

**Post-operative:**

Pain :

Oral Feeding:

Wound infection :

Frequency:

Others:

**Histopathology:**

**FOLLOW UP:**

Three months:

Six months:

One year:

There after:1.

2.

3.

Constipation / Incontinence:

Obstruction / Enterocolitis:

Rehospitalisation:

Additional Procedures:

<b>S.No</b>	<b>NAME</b>	<b>AGE</b>	<b>SEX</b>	<b>IP NO</b>	<b>TRANSITION ZONE</b>	<b>D.O.S</b>	<b>COMPLICATIONS</b>
1	SIVANESAN	2/12	M	49394	RECTO SIGMOID	19.05.06	
2	BALU	6/12	M	16565	RECTO SIGMOID	21.07.06	STRICTURE, ENTEROCOLITIS
3	B/O DHANALAKSHMI	27/365	F	49928	RECTO SIGMOID	18.10.06	STRICTURE
4	B/O SHANTHI	25/365	M	52753	RECTO SIGMOID	17/12.06	
5	CHITRA	2/12	F	67021	RECTUM	29.12.06	CONSTIPATION
6	SANFUR	1½	M	18006	RECTO SIGMOID	20.02.07	
7	B/O NAGARATHINAM	29/365	M	48992	RECTUM	03.04.07	DEATH
8	KRISHNA PRASAD	2	M	45210	RECTO SIGMOID	24.04.07	
9	SANTOSH KUMAR	5/12	M	39212	SIGMOID	08.05.07	STRICTURE
10	B/O MANJULA	4/365	M	18249	RECTO SIGMOID	25.05.07	STRICTURE

<b>S.No</b>	<b>NAME</b>	<b>AGE</b>	<b>SEX</b>	<b>IP NO</b>	<b>TRANSITION ZONE</b>	<b>D.O.S</b>	<b>COMPLICATIONS</b>
11	MANOJ KUMAR	4	M	65074	RECTUM	06.06.07	STRICTURE
12	SANJAY	2	M	66802	RECTO SIGMOID	31.06.07	
13	B/O KALAIMANI	19/365	M	25730	RECTUM	21.07.07	
14	HARISH	2/12	M	35378	RECTUM	12.08.07	CONSTIPATION
15	SANJU	5/12	M	66802	RECTO SIGMOID	30.08.07	ENTEROCOLITIS
16	B/O RADHA	2/12	M	29949	RECTUM	04.10.07	SOILING
17	B/O JOTHI	19/365	M	56390	RECTO SIGMOID	18.11.07	
18	KRISHNA	3/12	M	48276	RECTO SIGMOID	11.12.07	DEATH
19	MAHAVAISANAVI	3	F	74445	RECTUM	25.12.07	CONSTIPATION
20	B/O KAVITHA	7/12	M	14171	RECTO SIGMOID	25.01.08	SOILING
<b>S.No</b>	<b>NAME</b>	<b>AGE</b>	<b>SEX</b>	<b>IP NO</b>	<b>TRANSITION ZONE</b>	<b>D.O.S</b>	<b>COMPLICATIONS</b>

21	B/O KRISHNAVENI	5/365	M	68861	RECTUM	29.01.08	
22	SATHYAN	6	M	10471	RECTO SIGMOID	19.03.08	
23	B/O AMUDHA	10/365	M	28058	RECTUM	21.03.08	
24	GOWTHAM	2	M	76170	RECTUM	28.03.08	STRICTURE
25	TAMILSELVAN	11/12	M	43426	RECTO SIGMOID	16.05.08	SOILING
26	B/O BANU	3/365	M	52986	RECTO SIGMOID	24.06.08	
27	JAYARAJ	1½	M	13020	RECTO SIGMOID	01.07.08	
28	PRIYA	6/12	F	57342	RECTO SIGMOID	29.08.08	
29	B/O JOTHIMANI	26/365	M	48922	RECTO SIGMOID	17.10.08	
30	MADAN KUMAR	1½	M	57022	RECTO SIGMOID	09.01.09	