Transanal Endorectal Swenson Pullthrough :Single Center Experience

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ABSTRACT:

BACKGROUND:

Hirschsprung's disease (HD) is a common cause of intestinal obstruction in children. Definitive treatments consist of excision of the aganglionic segment and anastomosing the normal colon to the anal remnant by trans anal swenson approach.

OBJECTIVE:

The purpose of this study was to evaluate the feasibility and safety of TESPT technique for management of HD & its results.

PATIENTS AND METHODS:

It is aprospetive study ,where 33children (30 boys and 3 girls) with proven HD were treated at our pediatric surgical department between April 2009 and November 2013. Their ages ranged from 6 months to 8 years. The case files were evaluated for age at operation, associated congenital anomalies, operative time, intraoperative blood loss and blood transfusion, time to resume oral feeding, hospital stay, postoperative complications, and postoperative continence state. The median follow-up period was 12 months(range 3 to 21 months). The diagnosis was confirmed by barium enema &/or by partial thickness rectal biopsy for histopathological examination for all cases. Most patients had a well-defined transition zone at or distal to midsigmoid colon on preoperative contrast enema

RESULTS:

Male to female ratio was 10:1. one patient had positive family history for Hirschsprung's disease . The definitive TESPT was done in the patients at a median age of 10 months(range 6 months - 8 years). Seven patients had primary pull-through without colostomy. The median operative time was 120minutes (range 90-150 minutes). The length of the resected bowel varied between 20cm and 42cm. The length of hospital stay was 3 to 5 days. The time to resume oral feeding was ranging from 24-48 hours. There are 4 patients with recurrence of obstructive symptoms (retained piece) . 3 patients(9.09%) required readmission for treatment of enterocolitis. Postoperative adhesive intestinal obstruction occurred in 1patient (3.03%). Soiling occurred in 3 patient out of 33pts.(9.09%). Perianal excoriation occurring in 7 patients(21.21%) ,1 patient(3.03%) was died. **CONCLUSION:**

Tespt technique is both feasible and safe technique in properly selected children with rectosigmoid HD in all ages. The technique is easily learned and is associated with excellent clinical results. **KEY WORDS:** hirschsprung's disease(HD),trans anal endorectal Swenson pull-through(<u>TESPT</u>)

INTRODUCTION:

Hirschsprung's disease (HD) is one of the most common surgical diseases in the paediatric age group with an incidence of approximately 1 in 5,000 live births^{.(1,35)} It is a congenital condition characterized by the absence of parasympathetic ganglion cells in the submucosal (Meissner's) and myenteric (Auerbach's) plexuses of the distal bowel. This leads to a functional intestinal obstruction presenting with delayed passage of meconium in the newborn or recurrent

constipation in the older child with failure to thrive. The aganglionosis usually involves the anus and a variable portions of the large bowel

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Rarely, the disease extends to involve the small bowel or even the whole of the intestine.^(2,35) Some patients may present with enterocolitisrelated diarrhoea and many untreated cases of HD usually die in their infancy from this complication.^(3,35) Barium enema may help with diagnosis but the classical transition zone may not be obvious in the first three months:^(4,35)Rectal biopsy remains the gold standard in confirming HD and it shows absence of ganglion cells and presence of hypertrophied nerve fibers.

Traditionally, surgical therapy for Hirschsprung disease has consisted of a proximal defunctioning colostomy, followed months later by a definitive reconstructive "pull-through" procedure in which

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the aganglionic colon is resected and the normally innervated bowel is brought down and sutured to the area just above the anal sphincter. Although many types of pull-through procedures have been described, the most commonly performed operations in North America have been the Duhamel. Swenson. and Soave (or endorectalpullthrough) procedures. In the past several decades, increasing numbers of pediatric surgeons have abandoned the routine use of a colostomy in favor of a 1-stage pullthrough, with multiple studies suggesting that this approach is safe and efficacious.^(5-,6,37) Over the past few years, the popularity of minimal-access surgical techniques has led to a number of modifications to the standard 1-stage operations. Georgeson described a laparoscopic approach that has been adopted by many surgeons and that is associated with less pain and a shorter hospitalization than standard open procedures.^{7,37}Subsequently, a number of authors described a solely transanal approach, which appears to be associated with the same advantages without the need for any intraabdominal dissection. Transanal procedures leave No scars; have less post-operative pain, and a shorter Hospital stay. The most commonly used technique for The transanal pull-through is endorectal dissection, Which leaves a long muscular cuff. which is usually split Posteriorly.^(8,9) It is well known that the long muscular Cuff that is left behind causes obstruction. A few studies In the literature have shown that The problem of the remnant rectal cuff can be avoided if the transanal resection of the aganglionic segment is performed in the manner described by Swenson; by dissecting the full thickness of the rectum.⁽⁹⁾ The purpose of this study was to evaluate the feasibility and safety of TESPT for management of HD & its results.

PATIENTS AND METHODS:

It is a retrospective study, where 33 children (30 boys and 3 girls) with proven HD were treated at our pediatric surgical department between April 2009 and November 2013. Their ages ranged from 6 months to 8 years .The case files were evaluated for age at operation, associated congenital anomalies. operative time. intraoperative blood loss and blood transfusion, time to resume oral feeding, hospital stay, postoperative complications, and postoperative continence state. The median follow-up period was 12 months(range 3 to 21 months). The diagnosis was confirmed by barium enema &/or partial by thickness rectal biopsy for histopathological examination for all cases. Most patients had a well-defined transition zone at or distal to midsigmoid colon on preoperative contrast enema.Likewise, infants who could not be successfully decompressed by rectal irrigation were considered not suitable for 1-stage TESPT. The cases that presented with enterocolitis (n=7), were treated first with rectal washout and third generation cephalosporins. Preoperative bowel preparation started 48 hours before surgery with rectal irrigation using normal saline every 12 hours either through the stoma (if on colostomy) or via the rectum in patients undergoing primary pull-through, in patients who had fecal impaction or solid stool, use oral polyethylglycol powder dilution in water or juice. after Oral & ABs(metronidazole erythromycin or trimethoprim) also preoperative used in preparation of these patients 48hrs before surgery. The patients were kept on non-residue diet 24 hours before surgery. Third generation cephalosporin was administered at induction of anesthesia and continued for 3 days after operation.



Figure 1: Contrast enema for Hirschsprung's patient.

Surgical Technique:

After induction of anesthesia, the patient is placed supine with the pelvis elevated at the end of the operating table with the lower limbs attached to a bar. The abdomen and perineum are prepared in the standard fashion. some surgeons prefer the (prone jack knife position) for assuming more direct access to mesenteric vessels. The perineum is prepared in the standard fashion. A bladder catheter is inserted. The anal canal is exposed with Scott anal or Lonstar retractor or with stay sutures (hitch suture). A circumferential row of 4-0 silk stay sutures is inserted approximately 0.5 to 1 cm above the dentate line. At the beginning ,a full thickness incision was made on the rectal wall posteriorly as no important

pelvic structures were encountered in that location. 0.5 cm above the dentate line just distal the traction sutures and lifted to circumferentially. The incision was extended all around the circumference, incising the full thickness of the rectal wall. Rectal mobilization was done by working on the surface of the rectal wall using fine diathermy needle to develop the plane . The traction on anorectal tube facilitates proximal extension of anorectal tube dissection until the level proximal to peritoneal reflection (approximately 10 to 15 cm above the dentate line) the dissection could be performed easily once peritoneal reflection was reached, allowing mobilisation of the rectum and sigmoid colon out of the anus without leaving behind any muscular cuff. Mobilization of the colon is continued as proximal to the grossly obvious or histologically confirmed transition zone as possible by dividing the rectosigmoid vessels after cauterizing them ligatures may required. In cases, where the macroscopic transition zone was visible, we removed an extra 5 cm segment of the dilated colon to ensure pulling through of the ganglionated segment of the colon. In doubtful cases, where there was no visible differentiation of the transition zone, a frozen section confirmation was sought. With the limited availability of the resources, this seemed to be the logical course of action .After resection of the aganglionic segment, the normally innervated bowel is pulled through and anastomosed to the remaining part above the dentate line using 4-0 slowly absorbable suture material (coloanal anastomosis). Feeding was allowed 24-48 hours the procedure. First rectal digital after examination was performed after 14 days. Routine anal dilations were performed once or twice weekly for at least 3 weeks. Follow-up was

arranged once weekly for 3 weeks followed by once monthly for 3 months then every 3 months thereafter.

RESULTS:

Male to female ratio was 10:1. one patient(3.03%) had positive family history for Hirschsprung's disease. 2 patients(6.06%) Was preterm. The most common clinical manifestation of the studied group are presented in tables (1) and their age at operation are presented in figure (1).

Associated anomalies are presented in table (2). We confirmed the diagnosis by barium enema & rectal biopsy in all cases. The transition zone was present at mid sigmoid in 6 cases, at recto sigmoid junction in 22 cases,& at rectum in 5 cases. 26 patients had colostomy prior to the pull-through , the colostomies were done in the right transverse colon. 19 of the colostomies were loop while 8 were divided colostomy & this shown in table (3,4). Complications of colostomy were prolapse occurred in 4 Pts. (15.3%) and wound breakdown that occurred in 1 Pt. (3.8%). The duration of the colostomy before definitive TESPT varied from 1 to 18 months with a median duration of 4 months before definitive TESPT.

The definitive TESPT was done in the patients at a median age of 10 months with a range of 6 months to 8 years. six patients had primary pullthrough without colostomy. Intra-operative blood loss was minimal and no blood transfusion was needed. The median operative time was 120minutes (range,90-150) minutes. The length of the resected bowel varied between 20cm and 42cm. The length of hospital stay was 3 to 5 days. The time to resume oral feeding was ranging from 24-48 hours. The first per rectal examination was performed 2 weeks after operation with the little finger and only 2 patients required repeated dilatations due to anastomotic stricture. There are four patients with recurrence of obstructive symptoms (retained piece) because of the transitional zone was not clear during dissection the colon& the frozen section biopsy was not available during operation.

three patients required readmission for treatment of enterocolitis (admitted after operation; 2 months, 4 months and 10 months). They were treated with saline rectal irrigation, no oral feeding and 3rd generation cephalosporins. Postoperative adhesive intestinal obstruction occurred in 1patient(3.03%). Soiling occurred in 3 patients(9.09%). Perianal excoriation was the commonest postoperative complications occurring in 7 of cases and resolved within 8-14 weeks postoperatively, one patient was died suddenly after 2monthes of operation??. there are no patient

need for colostomy secondary to anastomotic leak.no patient had prolapse of the pulled through colon, these complications are presented in table(5).



Figure 1: Age distribution of patients at definitive Swenson Procedure.

Clinical features	NO.	(%)
Delayed passage of	30	90.9
meconium		
Chronic constipation	22	66.7
Abdominal distension	21	63.6
Enterocolitis	7	21.2
Vomiting	5	15.1
Faecaloma	4	12.1
Failure to thrive	1	3
Caecal Perforation	1	3

Table 1: Clinical presentation in children with HD that had pull-through operations.

Table 2 :Associated anomalies.

	NO.	%
Down`s syndrom	1	3.03
Hypospadius	2	6.06
Cerebral palsy	1	3.03

Table 3	:	Site	of	transitional	zone
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	NO.	%
Mid sigmoid	6	18.2
Recto sigmoid	22	66.6
Rectum	5	15.1

Table 4 : Types of TESPT according to number of stages.

Types of TESPT	TYPE OF COLOSTOMY	NO.	%
One stage		6	18.2
Two stages	#		
Three stages	27	81.8	
Divided colostomy			24.2
Loop colostomy	19	57.5	

#. We will need frozen section biopsy (not available) to determine the most distal ganglionic part of bowel(site of colostomy in two stages type)

Complications of TERSPT	NO.	%
Postoperative adhesive intestinal obstruction	1	3.03
Peri-anal excoriation	2	6.1
Recurrent constipation from residual aganglionosis	5	15.1
Anastomotic stricture	1	3.03
Post operative enterocolitis	3	9.09
Soiling	3	9.09
Death	1	3.03
Recto urethral, recto vaginal fistula		
Anastomosic leak		
Rectal prolapse		

Table 5 : Complications.

Study	No. of pts. ^{\$}	Pre op. Dx ^{\$\$.}	Frozen section	Leak	EC***	STRICTURE	Dilatations	OT** (minutes)
Gao et al., 2001	33	Barium enema, Manometry, rectal biopsy	yes	none	6.06%	3.03	yes	160 (85 – 260)
Weidner et al; 2003	15	Rectal biopsy	yes	none	13%	NA*	yes	158 (110 – 190)
Peterlini et al., 2003	22	Rectal biopsy	yes	9%	nil	NA*	no	NA*
Zhi – lin et al. 2008	134	Barium enema or Rectal biopsy	yes	none	2.9%	NA*	NA*	70 (50 – 115)
Sookpotarom et al; 2009	27	Barium enema or Rectal biopsy	no	none	11.1%	22.2	yes	153 (60 – 400)
Mahjan et al 2010	17	Clinical features and Barium enema	Yes (7pts.)	none	11.7%	11.7%	Yes (2 pt)	141 (120 – 200)
Swande et al 2011	33	Barium enema ,rectal biopsy	yes	3.03%	6.1%	3.03%	yes	NA*
Present study 2014	33	Barium enema ,rectal biopsy	no	none	9.09%	3.03%	yes	120 (90-150)

Table 5 : Comparison of various studies of TESPT.

*NA : not available

**OT;operative time

***EC;enterocolitis

\$ NO.of pts.;number of patients

\$\$ Pre op. Dx.;pre operative diagnosis

DISCUSSION:

Ninety five percent of full term infants pass meconium in the first 24 hours of life but less than 10% of children with HD pass meconium during that time. (10) in our study, 90.9% of cases experienced delayed passage of meconium. Seventy five% of patients with HD will demonstrate a transition zone on barium enema. The absence of transitional zone doesn't rule out the diagnosis.^(11,38). The first reconstructive operation for Hirschsprung's disease was described by Swenson and Bill in 1948.^{8,9} Since then, various other trans-abdominal pull-through procedures like Soave, Duhamel, and Rehbein have been described. The first report on the transanal procedures for the classic recto-sigmoid HD was published in 1998.^(12,9) The primary transanal pullthrough techniques with various modifications have become the standard of care around the world, each having their own proponents.^(13,9) The advantages of one-stage transanal procedures are numerous, ranging from the avoidance of multiple laparotomies and the preliminary colostomy with their associated complications, reduced operating time, less blood loss, no pelvic structure damage, single hospital admission, short hospital stay, decreased need for analgesics, significantly lower hospital costs, and no external scars, and with improved cosmetic appearance. ^(9,14-15) without compromising on the functional outcomes Above all, there is a high degree of parental acceptance of the procedure, as it avoids the need to care for a baby with a colostomy.^(9,16)

However, the aganglionic rectal muscular sleeve in the transanal Soave's procedure limits the volume of the replaced sigmoid and the rectum.^(17,9) This remnant rectal cuff also causes cuff strictures, diarrhoea, and incontinence.^(18,9,19) Many authors have recommended that this cuff be divided down to the sphincter to avoid constriction of the pulled through bowel, but this has remained controversial. (9,20) Falchetti et al reported that the cuff can roll down during the transanal pull-through and form a constricting ring just above the anastomosis.^(9,21) To avoid this problem, a few authors have used a much shorter mucosectomy with a shorter muscle cuff measuring only 1-2 cm -above the dentate line.^(13,9,21) Gao et al. described primary transanal recto-sigmoidectomy with the potential advantages of a partial rectal mucosectomy, internal shorter muscular cuff. anal sphincterotomy, and oblique anastomosis. (9,22) A few authors have entirely eliminated the submucous dissection and described a transanal Swenson procedure without any residual muscular cuff.^(23,9,24)

We have performed a transanal Swenson's procedure in selected cases where the preoperative contrast enema showed a distinct transition zone, with dilatation of the proximal normal bowel. The combination of the clinical history , the contrast enema findings and preoperative rectal biopsy was 100% accurate for the diagnosis. Preoperative rectal biopsy makes the transanal dissection difficult due to scarring and fibrosis, which may affect the functional outcome^(25,9) The location of the transition zone, as seen on contrast enema, has correlated accurately with the anatomical changes in the bowel in 28 out of 33 pts. (84.8%) of our patients. The loss of the anatomical transition zone in some of the patients can be due to the use preoperative rectal wash outs in small age group or due to presence of colostomy in case of multiple stages procedures . Procter et al. had shown that the radiographic transition zone correlates accurately with the level of aganglionosis in 90% cases of recto-sigmoid HD.^(9,26) In 28 Out of 33pts.(84.8%) patients in whom there was a classical anatomical transition zone, we could successfully avoid the frozen section biopsy, thus reducing the operating time. This becomes very important at the centres where the facility for frozen section is either unavailable or not well standardised. Sookpotarom et al. performed the transanal Swenson's procedure in 27 patients, based only on the radiological findings and intra-operative assessment of the anatomical changes, without the use of the frozen section technique.

Their assessme showed an accurate correlation with the final histopathological level of the aganglionosis in 85% of the patients.^(27,9) In the present study There was no need to taper the dilated bowel. We did not do an oblique anastomosis.^(28,9) but there are post operative obstructive problems in 5 pts out of 33(15%).

With strict dissection on the rectal wall, injury to the urogenital tract and the pelvic nerves could also be avoided.

A comparison of the various studies, in literature, for transanal Swenson's procedure is shown in Table 5.^(9,36) Continence has been generally satisfactory in most patients in this study. The incidence of faecal incontinence varies from less than 1% to as high as 32.7%.^(29,30,36) Although we do not have facilities such as anorectal manometry for measuring sphincter function in these patients clinical history and examination revealed that most of our patients are continent, where Three of them complained of soiling following surgery. all of them were had just minor soiling which improved over time. None of the patients in our study developed urinary

incontinence. Strict adherence to operative technique will avert this possibility in majority of the patients.

The anastomotic leak rate in this study was 0% which was comparable to that (5.6%) reported by Sherman et al., in larger series.^{31,36}, that may be related to good vascularity and good surgical techniques in combination with adequate bowel preparation, which are very important in any bowel anastomosis . In an era where colostomy for HD is being avoided because of its associated morbidity and even mortality, but in our study, there are 26 out of 33 of our patients required preliminary colostomy as part of staged management, The reasons for this colostomy were due to late presentation in the patients which makes a primary pullthrough unsuitable because of the gross bowel distension. Also, many of the colostomies were done as a life-saving procedure in some of them as the conditions of the patients precluded any attempt at a primary extensive surgery. The advantages of a preliminary colostomy are numerous and include relief of obstruction, time for dilated colon to regain caliber and tone, making for easy colo-anal anastomosis as well as easy bowel washout prior to the pull-through. It may also help to reduce the incidence of postoperative enterocolitis. Planning a two-staged procedure requires that the stoma be well sited in a biopsy-proven normal bowel (most distal ganglionic part of bowel), This facility was not constantly available during the study period . one stage TESPT was done for 5 out of 33 patients(15.2%), Those 5 patients need to be carefully selected in addition to adequate bowel preparations. The authors did not find a high incidence of postoperative enterocolitis in our patients following TESPT 3patients (9.09%) of our patients had suspected enterocolitis based on clinical presentation which responded to intravenous fluids and antibiotics .

The adhesive small bowel obstruction after open pull-through for HD ranged from 5% to 10%.^(32,33,36) This is comparable to what was obtained in this study1 patient (3.03%). The operative mortality associated with TESPT is low in many studies and ranges from 0-2.5%.^(31,34,35,36) In this series there was one death, (sudden death after 2 months of operation) giving a rate of 3.03%, autopsy of this patient was not available.

a redo transanal Swenson's operation was necessary in 5 patients (15.1%) who had residual aganglionosis. This was accomplished without difficulty and with good results.

CONCLUSION: AND RECOMMENDATIONS:

Transanal Swenson procedure is a feasible and safe undertaking in the properly selected cases of HD in all ages . The technique is easily learned and is associated with excellent clinical results, It not only avoids multiple abdominal operations, but also avoids the problems associated with the long muscular cuff of the transanal Soave's procedure. By adhering to the principle of dissecting on the rectal wall, injury to the surrounding structures and voiding disturbances of the bladder can also be avoided. This approach can be used safely in the patients of recto-sigmoid aganglionosis(short segment aganglionosis).

Both the transitional zone and a part of the dilated colon proximal to it should be excised to decrease risk of post operative constipation.

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