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Research Article

Trans abdominal Soave's-Boley Endorectal pull through for Hirschsprung's disease: A retrospective analysis of 57 cases

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ABSTRACT

Introduction: Hirschsprung's disease is a congenital disease of enteric nervous system in which ganglion cells fail to migrate through neural crest and results in absence of ganglion cells in submucosal and myenteric plexus and lead to a ganglionic intestinal segment which causes intestinal obstruction. Different surgical techniques are being used till date. Swenson's procedure, Soave-Boleyendorectal pull-through, Duhamel's procedure and transanalendorectal pull-through are most comnly used procedures which are being used now with certain modifications. Latest of all is single incision laproscopic surgery being used to compliment the transanalendorectal pull-through and Duhamel's procedure.

Purpose: The aim of the study was to evaluate the results of Soave's- Boley transabdominal pull-through in patients of Hirschsprung's disease in resource limited centers or peripheral hospitals.

Patients and Methods: It is a retrospective study of 57 patients,presenting to pediatric surgery department of DHQ teaching hospital Sahiwal, which were diagnosed withHirschsprung' disease who underwent Transabdominal Soave's-BoleyEndorectalpullthrough in the period of 10 years(dec 2007- dec 2017). In this study,there were 42 males and 15 females. Out of 57 patients , 52 were having short segment disease while 5 of them have long segment disease. Out of these long segment disease patients 2 of them were having total colonic aganglionosis.

Results: Out of 57 patients, most common complication was enterocolitis which developed in 15 cases(26.3%). Other complications were; wound infection in 05(8.7%), constipation in 04(7%), stricture in 02(3.5%), Incontinence in 01(1.75%) and no cases of anatomic leak or overflow incontinence were reported.

Conclusion: While in most centers in the developed world, Endorectal pull through operation for Hirschsprung' disease have evolved from open technique through laproscopically assisted pull through to totally transanal technique. However the open staged Soave-Boleyendorectal pull through procedure still has a role to play in the manangement of Hirschsprung's disease in resourse limited centers where facilities like laproscopic expertise and equipment, frozen section expertise and equipment, automatic stapler and neonatal intensive care units are not available.

Keywords: Hirschsprung's disease, Soave's-BoleyEndorectal pull-through, outcome

INTRODUCTION

Hirschsprung disease is the commonest cause of functional intestinal obstruction in children. It is a congenital malformation, characterized by absence of ganglion cells in submucosal

(Meissner's) and myenteric (Aurbach's) plexuses in distal bowel extending proximally for variable distances [1].Hirschsprung disease is caused by the failure of ganglion cells to migrate cephalocaudly through the neural crest during fourth to twelth weeks of gestation, causing an absence of ganglion cells in all parts of colon [2].Hirschsprung's disease is anomaly of enteric nervous system of neural crest origin so referred as neurocristopathy [3]Short segment disease is most common and is confined to rectosigmoid region of colon. Rarely babies are afflicted by near total intestine aganglinosis.

Its incidence is approximately 1/5000 birth and males are more frequently affected than females with ratio 4:1. Most of the patients present during neonatal period. Delayed passage of meconium beyond the first 24 hours is present in approximately 90%. Approximately 10% patients of Hirschsprung's disease present with Hirschsprung disease associated enterocolitis(HAEC).

Diagnosis of this disease is usually made during first year of life and rectal biopsy of mucosa and sub-mucosa is gold standard test for it.

Different congenital anomalies and syndromes like down syndrome(trisomy 21), neurocristopathysyndromes and congenital central hypoventilation syndrome may be associated with Hirschsprung' disease. It occurs as an isolated trait in 70% patients, as associated with chromosomal disorder in 12% cases, and with other congenital anomalies in 18% cases.[4-9]

Over the passage of time there has been a significant evolution in surgical strategies to treat Hirschsprung's disease. Since its first description by Harold Hirschsprung in 1896, various modalities of treatment have been described by many authors [10,11] but in 1948 Swenson *et al* [12] gave the basic principles of repair which further progress to two- or three- stage procedure to a primary operation in early 1980s. Three endorectal pull through techniques are used: Swenson, Soave and Duhamel. Further advancement came in its treatment in the form Laproscopic-assisted primary pull-through by Georgeson *et al* [13] in 1995. After this surgeons in the developed countries started shifting towards the laparoscopic procedure. Afterwards the entirely transanal endorectal pull- through emerged in 1998[14]. Most recent advancement is single incision laparoscopic surgery which is used safely to compliment the Transanalendorectal pull-through and the Duhamel's procedure.

Current regenerative strategies are under investigation to restore function in aganglionic intestine. Stem cell transplantation to regenerate the ENS is a subject of many recent experimental series[15].

PATIENTS AND METHODS

It is a retrospective study in 57 patients, presented to pediatric surgery department of DHQ teaching hospital Sahiwal with the diagnosis of Hirschsprung disease. All patients underwent trans abdominal Soave's-BoleyEndorectalpullthrough in the period of 10years (dec 2007- dec2017). There were total 57 cases, which were being operated by this procedure. In 55 patients endorectalpullthrough was performed withBoley,s modification of Soave,s procedure. In two patients ileoanalendorectalpullthrough with Boley,s medication was performed for total colonic aganglionosis. Out of 57 patients, 42 were males and 15 were females. All patients were diagnosed on the basis of clinical presentation and investigations. The sequence of investigations for making a definite diagnosis was, plain x ray abdomen, barium enema,full thickness rectal biopsy, laparotomy with serial segmental biopsies and colostomy/ileostomy. 39 patients presented in neonatal age group out of which 19 patients were presented with failure to pass meconium/intestinal obstruction and 20 patients presented with neonatal intestinal perforation.18

patients were presented beyond the age of 1 month.Out of 57 cases, 39 were diagnosed before 30 days of age, 05 between 30-89 days, 06 between 90-180 days and 07 beyond 180 days of age. Most of the patients were being operated at the age between 1-2 years(40 cases). 14 patients were operated between 2-3 years and 3 patients were operated between 3-4 years.Extent of disease was determined in all of these patients with the help of contrast enema, by naked eye appearance at laparotomy/colostomy and confirmed by serial segmental biopsies. From these patients , 52 were having short segment disease(29 uptorectosigmoid ,22 upto sigmoid colon and 1 uptodesending colon) while 5 of them have long segment disease. Out of these long segment disease patients, 03 patients were having disease upto transverse colon and 02 of them were having total colonic aganglionosis.

RESULTS

In these patients, most common complication was entercolitis which developed in 15 cases (26.3%). Other complications were; wound infection in 05(8.7%), constipation in 04(7%), stricture in 02(3.5%), Incontinence in 01(1.75%) and no cases of anatomic leak or overflow incontinence were reported. The colostomy/ileostomy was performed in all patients. The complications of stoma are shown in the table no . Recurrent prolapse of stoma and skin excoriation were most common complications.Skin excoriation was present in15 (26.3%) patients and was managed by local applications like petroleum jelly. Recurrent prolapse of stoma was a troublesome complication seen in 10(17.5%) patients. Episodes of severe diarrhea were encountered in patients with ileostomy,9(15.7%).Retraction of stoma was present in 2(3.5%) patients

Table 1: Gender Distribution

Sex	No. of patients
male	42
female	15

Table 2: Age distribution

Age of patients	No. of patients
< 30 days	39
30-89 days	05
90-180 days	06
> 180 days	07

Table 3: Age at the time of Operation

Age of patients	No. of patients
< 1 year	00
1-2 years	40
2-3 years	14
3-4 years	03
4-5 years	00
> 5 years	00

Table 4: Mode of Presentation

No. of patients	Presentation
19	Neonatal intestinal obstruction
20	Neonatal intestinal perforation
18	Distention and constipation in more than one month of age

Table 5: Level of aganglinosis

Level of aganglinosis	No. of patients
Recto sigmoid	29
Sigmoid Colon	22
Descending Colon	01
Transverse Colon	03
Ascending Colon	00
Total Colonic	02

Table 6: Complications (definitive procedure):

Complications	No. of patients
Wound infection	05
Anastomotic leak	00
Stricture	02
Constipation	04
Incontinence	01
Overflow incontinence	00
Enterocolitis	15

Table 7: Complications (stoma):

Complications	No. of patients
Skin excoriation	15
Stoma prolapse	10
Stoma retraction	02
Diarrhea	09

Table 8: Associated anomalies:

Anomalies	No. of patients
Down syndrome	02
Walden burg shah syndrome	01
Anorectal malformation	01
Left PUJ obstruction	01
Sensorineural hearing loss	01
Type 1 Diabetes mellitus	01
Hypothyroidism	01

DISCUSSION

Harald Hirschsprung described the gross anatomy of this disease. A lot of research work had to be done to understand the etiology and pathophysiology. Erenphresis 1946, was the first to point out that absence of ganglion cells is the basic cause of the hirschsprung's disease. The surgical management of hirschsprung' disease has gradually evolved overtime since the basic principles of repair described by Swenson et al . There are several surgical procedures that are used for the treatment of patients with hirschsprungdiasease[16]. Despite the advances in surgical techniques, there are still children with functional problems even after surgical treatment[17]. Three surgical procedures are known to offer satisfactory outcomes. Swenson 'scoloanal anastomosis, Soave's endorectal pull through and Duhamel's retrorectal pull through are being performed commonly. All these procedures are based on the principle of bringing normally innervated bowel down to the anus. The

difference is merely in the type of anastomosis and methods of preserving and using the aganglionic rectal pouch. The Swenson's procedure involves an end to end anastomosis after resecting aganglionic colon between normally ganglionic bowel and anus.Duhamel's procedure is an end to side anastomosis between the rectal pouch which is aganglionic and the normal ganglion bowel brought behind the rectal pouch and anastomosed to its posterior wall. Soave's procedure is a coloanal anastomosis between the normal ganglionic bowel pulled through a seromuscular cuff of the aganglionic rectum, prepared by stripping the mucosa. The laparoscopic assisted pullthrough procedures and entirely transanalpullthrough procedures are commonly performed in well developed centers. The functional outcome in patients of hirschsprung disease is variable markedly. In our case, all patients that presented with hirschsprung disease had trans abdominal Soave-boleyendorectal pull through after radiological and histological confirmation. This procedure does not require any special surgical gadgets like stapling gun. There is no pelvic dissection and there is no direct weakness of internal anal sphincter. This procedure has been widely practiced with a lesser complication rate,as shown by Kleinhaus et al (1979) and Ikeda and Goto (1984) that 60.4% and 64.9% of the patients treated with primary anastomosis at the time of endorectalpullthrough was free of complications respectively. The incidence of enterocolitis was also less with this procedure (2.1%) while it was 15.1% with Swenson, 5.9% in Duhamel and 5% with Soave's pullthrough. In our series it was noted in 15(26.3%) patients during first year of followup but latter on it dropped down to 5(8.7%) . The results of modified Soave's operation in our series are compareable with the results in literature [18,19], 89.25% of our patients are fully continent. Zain et al [20] studied Swenson and Soave pullthrough from june 2006 to june 2010. Complication rate after Swenson procedure was 24% while following soave procedure, it was 20%. Commonest complication after Swenson pullthrough technique were wound infection and adhesive intestinal obstruction(12%) while commonest complication after Soave procedure was anastomotic stricture(20%). In our study, commonest complication was enterocolitis(26.3%). Anastomotic stricture was present in(3.5%). A recent long term study has shown a significantly better continence for transabdominal approach compared with transanalpullthrough[21] Transabdominal single stage Soave pull through is safe, applicable and with lower associated complications in resource limited settings[22]. Benefits of minimally invasive surgery has been demonstrated in terms of shorter hospital stay and improved cosmesis and other potential benefits are hypothesized. Major improvements in functional outcomes remain as yet unproven[23]. The study shows the results of transabdominal Soave Boley ,spullthrough procedure are good and equally comparable with the results in literature as for as functional outcome is concerned.

CONCLUSION

While in most centers in the developed world, Endorectal pull through operation for Hirschsprung' disease have evolved from open technique through laproscopically assisted pull through to totally trans anal technique. However the open staged Soave-Boleyendorectal pull through procedure still has a role to play in the manangement of Hirschsprung disease in resourse limited centers where facilities like laproscopic expertise and equipment, frozen section expertise and equipment, automatic stapler and neonatal intensive care units are not available.

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