Hirschsprung’s disease management: From multi staged operation to single staged transanal pull-through

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ABSTRACT
Hirschsprung’s disease (HD) is a common congenital disease of colorectum. Although it was described more than one century ago, the effective treatment was established only half a century later. The initially treatment consisted of preoperative diverting colostomy, followed by definite pull-through and closure of colostomy on later date. A variety of procedures like Swenson, Duhamel, Rehbein and Soave were evolved with time. With the better understanding of pathogenesis and improvement on surgical technique, now the operation has become less extensive. In recent days, the classical 3 staged procedure is replaced by single staged procedure. All the procedure can be completed transanally. Since De la Torre in 1998, first reported total transanal endorectal pull-through, it became the most popular technique for the treatment of HD.

Keywords: Hirschspurg’s disease, multistage operation, single stage operation.

INTRODUCTION
Hirschspurg’s disease (HD) is one of the commonly studied diseases among pediatric surgeons and researchers. The clinical entity similar to HD was found in literature for more than 350 years. Description by Frederick Ruysch in 1691, probably, was the first known description of HD. The modern history of HD starts from 1986 when Dr. Harald Hirschsprung (1830-1966) presented a paper during a conference of the German Society of Pediatrics in Berlin (first published in 1988). He described 2 infants who died of complications of bowel obstruction. The large intestine was markedly dilated and hypertrophied whereas the rectum seems to be normal. After this many similar cases were reported including 10 more cases by himself in 1904.

However, pathogenesis of HD was not clear till next half century. The effective treatment modality could not be established because of ill understood pathogenesis. As early as 1901, Tittel found the absence of ganglion cells in the distal colon of HD, but significance could not be established at that time. In 1948, Ehrenpreis explained the present concept of pathogenesis of HD. Congenital absence of ganglion cells in distal colon causes functional obstruction and proximal dilatation is due to distal obstruction. In the same year, Swenson and Bill performed the first corrective surgery by removing the aganglionic colon and anastomosis. After this procedure, there have been a lot of modifications. Each is claimed to have less technical difficulties and better outcome. The main concept of all procedures is surgical removal of aganglionic segment or bypass. The aim of the article is to discuss the evolution of different surgical procedures, mainly multistage to single stage pull-through.

PRE SWENSON ERA
The initial attempt to cure the disease were a diverting colostomy by various authors. The procedure relieved the symptoms but recurred after closure of colostomy. This, now we can explain by persistence of disease in the distal colon. The procedure could not give permanent solution. However it became the established procedure as a temporary and rescue procedure. Subsequent procedures like Swenson, Duhamel and Soave were also preceded by a diverting colostomy. Different other procedures were also tried: procedures similar to posterior anorectal myoectomy, spinal anesthesia, sympathectomy and resection of different portion of colon. Treves was close to the definitive treatment. He treated a 5 year girl by excising dilated, narrowed segment of colon including entire rectum, anal canal and anus and creating perineal colostomy.

SWENSON PROCEDURE
In the same year 1948, Ehrenpreis explained the pathogenesis, Swenson and Bill performed the first successful corrective surgery. This was the historic breakthrough in the understanding of pathogenesis and treatment of HD. The procedure, soon became popular as Swenson’s procedure, brought a realistic hope that children with HD can be cured.
The procedure includes aganglionic rectosigmoidectomy with end to end anastomosis of normal colon just above the anal sphincter. After laparotomy, sigmoid colon is mobilized up to the peritoneal reflection. Then rectum is freed from perirectal tissues by gentle and careful dissection around the rectum down to the anal canal. The mobilized aganglionic rectum and anal canal is everted temporarily and anastomosis between the pulled through ganglionic colon and anal canal is performed outside anus. The level of anastomosis is about 2 cm above the dentate line. After anastomosis, it is reduced back to anal canal. There is a considerable risk of injury to pelvic nerve and blood vessels because of perirectal dissection.

The Swenson procedure is being performed for more than half century. In a recent follow up study of 2 decades, constipation was in 9.8% and incontinence in 3.1%. Follow up reports up to 40 years are available and showed that 96% were normal.

REHBEIN PROCEDURE

The description of Rehbein procedure was found in some literatures especially from some European countries. The newer text books do not mention this as a type of an operation. The principle of treatment of Rehbein method is removal of aganglionic colon up to upper rectum (about 2 cm from peritoneal reflection) followed by a vigorous dilatation of the remaining rectum and anal canal. According to Rehbein, the achalasia of anal canal is the most important cause of post operative obstruction, not the residual aganglionicosa at the anal canal. A special collagenous fibrous layer, adventitia rectalis, limits the free expansion of the rectal wall. Dilatation of anus is the most important part of the operation. Constipation is more commonly found in the created retrorectal space. Anastomosis is performed with the posterior wall of the native aganglionic rectum. Use of stapler is helpful to remove the common wall between the native rectum and the neorectum. GI continuity is maintained leaving a significant portion of aganglionic rectum. This requires much less pelvic dissection and preserves of the extrinsic innervations. The procedure is benefited by use of stapler. The abdominal dissection can be completed by use of laparoscopy also.

This procedure is relatively easy and preserves the nerves around the anus. Duhamel reported less mortality, constipation, diarrhea, anastomotic leak and intestinal obstruction. The possible disadvantages are anastomotic stricture and complications related to blind pouch and spur. Baillie et al studied the long term outcome of colonic motility after Duhamel procedure for HD. The outcome after the procedure was not up to the same level as a normal child. The satisfactory outcome was achieved only in 42%. A long term study by Zhang et al showed a satisfactory result but still it is not as expected in normal child soiling was in 27%, constipation in 9.8% and incontinence in 12%. The other short term and long term complications and outcome also varied from satisfactory to good.

SOAVE PROCEDURE

In 1964, Soave described the details of endorectal approach for pull through. The endorectal pull-through was originally described through transabdominal approach. The operation involves removal of rectal mucosa-submucosa and pulling through the normal ganglionic colon through the muscular cuff of aganglionic colon. Endorectal dissection starts 2 cm below the peritoneal reflection by incising the seromuscular layer circumferentially. The dissection in seromuscular plane is continued in distal direction up to 5 mm of dentate line. The mucosal-submucosal tube is everted and brought through anus and cut at 5 mm above dentate line. The normal ganglionic colon is brought to this position and coloanal anastomosis is done.

The concept of development of this method is to prevent extensive dissection at rectum. The preservation of normal muscular cuff also preserves the innervations around anal sphincter. In this procedure, the presence of aganglionic muscular cuff around normal ganglionic colon may increases the risk of constipation. But, fortunately, it is not seen clinically.

TRANSANAL MUCOSECTOMY

Rintala and Lindahl, first reported the transanal endorectal pull-through for HD in 1993. Rectum was
dissected caudally to the upper border of the levator muscles. Then mucosal proctectomy was performed transanally beginning 3-5 mm above the dentate line. Rectum was excised transanally so that 2-3 cm of muscular cuff remained. The ganglionic colon was pulled through the aganglionic muscular cuff and anastomosed at mucosal edge. They reported normal age appropriate bowel function in a follow up up to 4 years. 21 In 1996, Saltzman et al reported a similar and complete resection of rectum by transanal approach. Their result shows a less operating time and less post operative hospital stay. The outcome was similar with the standard transabdominal pull through.22

LAPAROSCOPIC PROCEDURE
Georgeson was first to describe laparoscopic approach for abdominal part of the pull-through.23 Their operation uses laparoscopy to identify the transitional zone, mobilize rectum below the peritoneal reflection and combined with a short mucosal dissection from transanal approach. The normal colon was then pulled down and anastomosed at anus 5-10 mm above dentate line. No patient had either preoperatively or post operatively colostomy. This has a clear advantage of avoidance of laparotomy. The procedure causes less stress and the outcome are equally good as compared with open procedure.24 Hospital stay and cosmetic results are superior in laparoscopy.25 Ishikawa found less operative complication while long term complication and outcome were quite similar.26 The additional advantage is clear visualization of perirectal structure even in small infants.

PRIMARY PULL-THROUGH
Till 1980s, almost all children used to have defunctioning colostomy at the time of diagnosis and definitive reconstructive surgery on later date. During the initial days, it was thought that it is unsafe to perform a definitive operation at neonatal period. Colostomy or ileostomy was the major initial step in the management of HD. The advantages are relieve of obstruction, less frequent enterocolitis and dilated non ganglionic bowel returns to normal size. The procedure gives time to improve nutrition. In 1980, So et al first described that diagnosis of HD does not always need a defunctioning colostomy.27 Constipation and mild enterocolitis can be adequately and safely managed by colonic irrigation. Primary endorectal pull-through without creating colostomy is also equally safe and has good results.28,29 Severe enterocolitis, perforation, malnutrition, massively dilated proximal bowel and inadequate pathological support to reliably identify the transition zone are still accepted indications for a prior colostomy.30 The third step of operation, closure of colostomy, can also be avoided if the initial defunctioning colostomy is closed during the definite pull through. There are only few reports available in literature on such types of 2 staged procedures.31

TOTAL TRANSANAL ENDORECTAL PULL-THROUGH
The most recent and popular procedure is the single staged total transanal pull-through (TTEP) developed by De la Torre and Ortega in 1998.32 This involves complete dissection and mobilization of entire aganglionic colon and anastomosing the normal colon through anus. No additional procedures like laparotomy or laparoscopy is required. The initial part of the surgical procedure is similar as described by Rintala and Saltzman. The dissection of mucosa submucosal tube extends proximally to reach peritoneal reflection. Then the muscular layer is opened and further dissection becomes full thickness. Once adequate mobilization of normal colon occurs the normal colon is brought into anus through the muscular tube and anastomosis done. Torre’s modification of Soave procedure is used by pediatric surgeon worldwide.33 This technique has various advantages over the other traditional procedures like Swenson, Duhamel and original Soave. This is minimally invasive, no abdominal incision results no abdominal scar and complications of laparotomy, cosmesis, preservation of sphincter, less operating time, less bleeding, less hospital stay, early recovery and less expenses.33-35

The outcome of any procedure for HD is measured as constipation and incontinence. The aim is to achieve a regular bowel movement without incontinence. The results from many papers are in favor of the single stage TTEP34-40 Constipation ranges from 0% to 8.6%,34,36,38,39 and incontinence from 0% to 17%.34,36,38,39 Huang et al compared the post operative bowel movement after TTEP with other types of procedures and found that the anal manometry parameter were similar.41 Jester et al has reported a higher rate of anastomotic dehiscence in 8 out of 34 patients. Interestingly, 6 of them were asymptomatic and detected during examination.42 The rate of dehiscence can be higher than our expectation. Early detection and resuturing can prevent the more serious sequele.

The redo endorectal pull-trough was also found to be feasible and safe irrespective the type of first procedure. Obermayr et al reported 8 patients with redo operation by endorectal pull-through. Out of them, 4 had normal bowel function and another 2 has only mild constipation.43
NEWER MODIFICATIONS

Further modification has been seen in many literatures. The standard primary transanal endorectal pull-through was modified by oblique rectal mucosectomy. The mucosectomy starts 1 cm posterior and 2-3 cm anterior making the posterior wall splits as ‘V’. This leaves the short muscular cuff and is claimed to have less post operative enterocolitis and without fecal incontinence and constipation.44

Hu et al described the dissection of rectal cuff in layers. It is similar to Soave to start with. Mucosa is cut at 5 mm above dentate line and dissection proceeds upward for 2 cm. Then circular muscle layer was carefully cut to expose longitudinal muscular layer. Longitudinal muscular coat is preserved for 1-2 cm and finally transected to enter the peritoneal cavity. This procedure leaves a thinner muscular cuff and claimed to have less post operative enterocolitis and constipation. However the dissection between layers is tedious.45

Sapin et al, described the transanal perirectal dissection. The procedure starts with a full thickness circumferential incision around 5-15 mm above dentate line and proceeds proximal in perirectal plane. The early and long term results were similar with endorectal procedure.46 Reports of similar procedures are also available from China.47 Xu et al describes the single stage transanal Swenson procedure with a reasonable outcome.48 Single staged fish mouth and parachute surgical technique is one of the modifications of Duhamel operation by Mousavi et al.49

In the last decade, after Torre’s modification of Soave procedure, a various modification has been seen. The common about all procedures is that the operations are single staged and performed totally through anus without need of laparotomy and laparoscopy.

HD is one of the important pediatric diseases, which carries a significant morbidity. The operative procedure is ever evolving. Earlier the operation used to be complicated and morbid procedure which requires multiple operations. In recent days, it has become simpler and less morbidity. The complete procedure shifted from 3 stages to single stage. Now, the entire procedure can be performed transanally. The outcomes of single stage TTEP are superior to other older procedures. The short term outcomes are encouraging. The outcomes when the child becomes adult are still waiting. Recent evidences prove this as a safe and effective procedure. Single stage primary TTEP can be considered as gold standard procedure of this time for the treatment of HD.

REFERENCES:


