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Ten-year experience with the Swenson procedure in Nigerian children with Hirschsprung's disease

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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. The outcome of this approach in Nigerian children is not known. The aim of this study was to analyze the short to mid-term outcome of children who have undergone the Swenson Procedure (SPT) as a treatment of HD over a period of 10 years. **Patients and Methods:** The clinical data of biopsy-proven cases of HD managed at the Obafemi Awolowo University Teaching Hospital, Ile-Ife, Nigeria, between 1998 and 2007 were reviewed.**Results:** There were 33 children 28 were males and five were females (M 5.6: F = 1). The median age at presentation was eight months (range three days to 11 years). Eighteen (54.5%) patients had primary pull-through without colostomy while 15 (45.5%) patients had a two-staged operation. The operation was carried out at a median age of 14 months with a range of seven weeks to 11 years, four months. Twenty-one (64%) of the patients were older than one year at the time of surgery. The level of aganglionosis was in the recto-sigmoid area in 30 (91%) patients. The commonest post-SPT complication was intestinal obstruction from adhesions. Bowel opening varied from once daily to eight times daily, median thrice daily. **Conclusion:** The Swenson's Procedure is an effective procedure in the treatment of HD in children in Ile Ife, Nigeria.

Key words: Children, Hirschsprung's disease, Nigeria, Swenson pull-through

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

Since its first description in by Harald Hirschsprung, the understanding and the management of the condition has improved greatly. The first successful treatment of the condition was undertaken over six decades ago by Orvar Swenson.[5] The original operation (the Swenson procedure) consisted of freeing the defective distal colon from within the pelvis by careful sharp extra-rectal dissection down to 2 cm above the dentate line and performing an end-to-end anastomosis. Since this first definitive operation was described, many other patients have been treated successfully with other operations including the retro-rectal pull-through of Duhamel and its modifications and the endorectal pull-through of Soave procedure and its various modifications.[6-9] Currently, these procedures are either laparoscopically assisted or accomplished completely via a trans-anal route without abdominal incision.[10,11]

There are few reports on the outcome of the surgical management of HD in our environment. Until recently we treated our patients with the trans-abdominal Swenson procedure. The aim of this study was to analyze the short to mid-term outcome of children who had undergone the original Swenson's Procedure (SPT) as a treatment of HD over a period of 10 years.

MATERIAL AND METHODS

Data were collected on biopsy-proven cases of HD who had SPT at the Obafemi Awolowo University Teaching Hospital, Ile Ife, Nigeria between January 1998 and December 2007. Information collected included age, sex and clinical presentation, type of colostomy done and complications if any, proximal level of aganglionosis, age at SPT, levels of resection, and any complication of SPT. Other information recorded included need for secondary surgery and bowel and sphincteric activities at follow-up. All the patients had plain abdominal X-rays but barium enema was done if the patient could afford it. Full-thickness rectal biopsy was done under general anaesthesia to confirm the diagnosis in all the patients. All patients had preoperative bowel washout either through the stoma (if on colostomy) or via the rectum in patients undergoing primary pull-through as well as intravenous peri-operative ceftriaxone and metronidazole. The operative technique is as described by Swenson and Bill with oblique anastomosis of the colon to the anal stump.[5] No deliberate attempts were made to add internal sphincterectomy. Patients undergoing primary SPT had frozen section biopsy to determine the presence of ganglion cells in the pulled through colon when possible. Frozen section biopsy was not consistently available during the period and level of resection is from a combination of barium studies' report, visualization of the transition zone and frozen section when available.

All anastomoses are either covered with a caecostomy tube using size 24-26 Foley's catheter or well lubricated 2-cc syringe inserted into the anus beyond the anastomosis which is removed on the fifth postoperative day. A gentle rectal examination to assess the anastomosis is done on the 10th day unless there is evidence of peritonitis. During follow-up, the parents were asked about the pattern of bowel motion, presence of continuous soiling and straining at defecation. The older children were also asked about rectal sensation as well as soiling.

RESULTS

There were 33 children managed with Swenson operations during the period, of which 28 were males and five were females (M 5.6: F = 1). The median age at presentation was eight months (range three days to 11 years). The clinical presentation in the patients was as shown in Table 1. Chronic constipation and abdominal distension were the commonest presentations. Despite the late presentation a history of delayed passage of meconium in the neonatal period was obtainable in 20 (60.6%) of the patients. All the patients had features of intestinal obstruction on plain abdominal X-rays while one neonate with caecal perforation had free intra-peritoneal gas. Barium enema was done on nine patients and classically demonstrated the transition zone.

Fifteen (45.5%) patients had colostomy prior to the pull-through. Eleven (73.3%) of the colostomies were sited in the sigmoid colon and the remaining four (26.7%) in the right transverse colon. Nine (60%) of the colostomies were divided while six (40%) were loop colostomy. The median age at formation of colostomy was six months (three days to 11 years). Major complications of prolapse and wound breakdown occurred in four (26.7%) and two (13.3%) of the patients with colostomy respectively. A patient had non-functioning stoma because it was sited blindly in an aganglionic segment. This required refashioning of the stoma to a more proximal level. The duration of the colostomy before definitive pull-through varied from one to 18 months with a median duration of four months.

The definitive SPT was done in the patients at a median age of 14 months with a range of seven weeks to 11 years, four months. Eighteen (54.5%) patients had primary pull-through without colostomy. There were 15 males and three females with age range from 15 days to 11 years. Bowel decompression by warm saline enema was used for a period of one to two weeks before pull-through. Majority (63.6%) of the patients were operated on after one year. The age distribution of the patients at pull-through was as shown in Figure 1; 90.9% of the patients had aganglionosis restricted to the rectosigmoid area as shown in Table 2.

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

Clinical features	Frequency (%)
Delayed passage of meconium	20 (60.6)
Chronic constipation	22 (66.7)
Abdominal distension	21 (63.6)
Vomiting	5 (15.1)
Faecaloma	4 (12.1)
Enterocolitis	3 (9.1)
Failure to thrive	1 (3)
Caecal Perforation	1 (3)

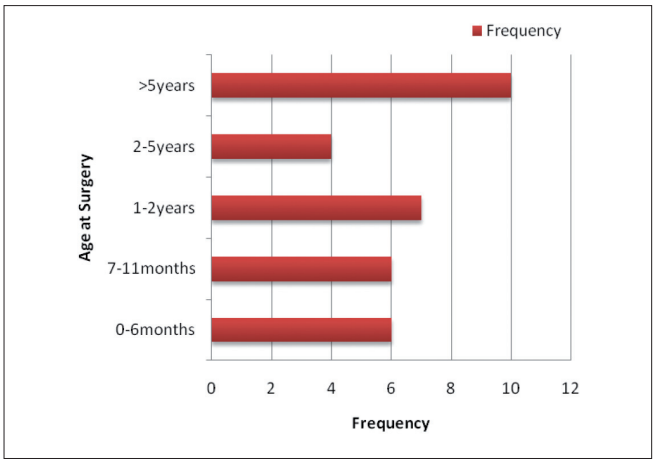


Figure 1: Age distribution of patients at definitive Swenson Procedure

One patient, an eight-month-old male infant died in the immediate postoperative period but the cause of death could not be ascertained as no authopsy was performed. He had a two-staged operation. Postoperative complications were as listed in Table 3. The commonest post-SPT complication was intestinal obstruction from adhesions which was managed conservatively in two patients while the third patient required laparotomy. Two other patients required laparotomy for drainage of pelvic abscess following anastomotic leak. One of them later required a posterior sagittal approach to restore colo-anal continuity. One patient who had primary pull-through required a redo because of residual aganglionosis. The duration of hospitalization following pull-through ranged from seven days to 44 days (median 11 days). Duration of follow-up ranged from four months to seven years (median six months).

In 30 (90.1%) of the patients, there was continence to flatus and faeces. The stooling frequency varied from once daily to eight times daily (median three times daily). There were two (6.1%) cases of occasional soiling which improved over time while there was one (3%) case of night-time incontinence. This patient had a staged procedure. One of our patients had suspected enterocolitis based on clinical presentation which responded to intravenous fluids and antibiotics.

Table 2: The level of aganglionosis as determined by barium enema and histology

Proximal level of aganglionosis	Frequency (%)
Rectum	12 (36.4)
Sigmoid colon	18 (54.6)
Descending colon	1 (3)
Transverse colon	1 (3)
Splenic flexure	1 (3)
Total	33 (100)

DISCUSSION

This study evaluated the outcome of the original Swenson operation for the treatment of HD in Nigerian children. Since Swenson described his original operation in 1948, there have been several modifications of techniques. Currently, there appears to be a shift towards transanal pullthrough as this is associated with less morbidity than open surgery. However, the choice of technique is still influenced by the training of the surgeon and the available resources and the presentation of the patient. At our hospital we have been using the open Swenson technique on all our patients with HD.

The results of this study suggest that the original Swenson operation still has a role to play in the management of HD in children, especially in our environment. The majority of our patients had a good postoperative outcome both in the short and medium term. Normal continence and regular bowel emptying is the primary goal in the surgery for HD. The dreaded possibility of urinary and faecal incontinency which led to several modifications of the technique did not surface in our patients. Continence has been generally satisfactory in most patients in this series. The incidence of faecal incontinence varies from less than 1% to as high as 32.7%.[12-14] 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. Three of the patients complained of soiling following surgery. In two of them there was just minor soiling which improved over time while one had major soiling mainly at night. None of the patients in our series developed urinary incontinence. Strict adherence to operative technique will avert this possibility in majority of the patients.

The anastomotic leak rate in this series was 6.1% which was comparable to that (5.6%) reported by Sherman *et al*., in larger series.[15] These leaks occurred in the initial phase of this study when a single-layered

Table 3: Postoperative complications

Complications of Swenson's pull-through	Frequency (%)
Postoperative adhesive intestinal obstruction	3(9.1)
Anastomotic leak with intra-abdominal abscess	2(6.1)
Wound infections /wound dehiscence	2(6.1)
Peri-anal excoriation	2(6.1)
Recto-vaginal fistula	1(3)
Recurrent constipation from residual aganglionosis	1(3)
Anastomotic stenosis	1(3)
Death	1(3)

anastomosis was done. Since we have modified our technique to using a double-layered anastomosis and in combination with the use of a rectal syringe tube vent we have not had a case of leak. We believe that the rectal vent decompresses the proximal colon preventing vigorous peristalsis against the anastomosis. It is generally accepted however, that good vascularity and good surgical techniques in combination with adequate bowel preparation 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, almost half of the patients required preliminary colostomy as part of staged management. The reason for this is 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 calibre 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. This facility was not constantly available during the study period due to cost constraints, mechanical failures of the frozen section machine as well as the emergency nature of the colostomy. Reliance had to be made in some cases on visual identification of the transition zone as well as presence of peristalsis on stimulation in order to site the stoma. One patient required resiting of the stoma because it was sited in the aganglionic portion. In a previous study we have shown that primary pull-through is feasible and desirable in our children even in infants less than four months old.[16] The patients, however, need to be carefully selected in addition to adequate bowel preparations. We were able to accomplish these in 54.5% of the children. The authors did not find a high incidence of postoperative enterocolitis in our patients following Swenson's PT. One of our patients had suspected enterocolitis based on clinical presentation which responded to intravenous fluids and antibiotics.

The operative mortality associated with SPT is low in many series and ranges from 0-2.5%.[15,17-19] In this series there was one death directly linked to the pull-through procedure giving a rate of 3.03%. The adhesive small bowel obstruction after open pull-through for HD ranged from 5% to 10%.[20,21] This is comparable to what was obtained in this series.

Sherman *et al*., reported that in 880 Swenson procedure cases the secondary re-operation rate was 6%. This stemmed from an anastomotic leak rate of 5.6%, which required diverting colostomies in most cases, and, in rare cases, the need to surgically excise strictures. Due to the inconstant use of frozen section biopsy facility during the study period, a redo Swenson's operation was necessary in one patient who had residual aganglionosis. This was accomplished without difficulty and with good results. Our series is however smaller compared to the 880 patients evaluated by Sherman *et al*.[15]

In conclusion, while in most centres in the developed world, pull-through operations for HD have evolved from the open technique through laparoscopically assisted pull-through to totally trans-anal pull techniques (TAPT), late presentation in our setting militate against this. It is hoped that with early presentation and diagnosis, TAPT would also become routine in our patients. However, the open Swenson procedure still has a role to play in the management of HD in our environment.

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