POST SURGICAL COMPLICATIONS OF HIRSCHSPRUNG'S DISEASE AND THEIR MANAGEMENT AT KENYATTA NATIONAL HOSPITAL: A 10-YEAR RETROSPECTIVE STUDY

BY

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DECLARATION

I, Francis Njoroge Kuria, do declare that this dissertation is my original work and has not been presented for a degree in any other University.

Signed .....

DR. F. N. KURIA

This Dissertation has been submitted for the degree of Master of Medicine (Surgery) with our approval.

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DEDICATION

This work is dedicated to my wife Jane and my children Jeniffer and Lilian.

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I would like to thank all those who made the completion of this study possible.

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SUMMARY

This is a retrospective study of the post-surgical complications of Hirschsprung's disease and their management at Kenyatta National Hospital during the period January 1991 to December 2000. Medical records of 96 patients who underwent surgery for histologically proven Hirschsprung's Disease at Kenyatta National Hospital and any associated colostomies closed were reviewed. Aganglionosis extended upto the rectosigmoid region in 75 (78.1%), proximal to the internal anal sphincter in 15 (15.6%) and proximal to the splenic flexure in 6 (6.3%)

The surgical procedures employed included Swenson's pull-through in 54 (56.3%), Soave-Boley endorectal pull-through in 39 (40.6%) and myectomy in 3 (3.1%). No patient had definitive surgery in the neonatal period, while 16 (16.7%) had surgery between one month and one year and 80 (83.3%) after one year of life.

Seventeen (17.7%) of the definitive procedures were one-stage, 17(17.7%), 2- stage and 62 (64.%) were 3-stage procedures.

Early post-operative complications occurred in 17(17.7%) and late complications in 70 (72.9%). The main complications observed were persistent constipation in 25 (26.0%), ano-rectal stenosis in 20 (20.8%), faecal incontinence in 21 (21.9%) and enterocolitis in 11(11.5%).

Anorectal stenosis was commonest after a Soave-Boley procedure (55%) compared with Swenson's (45%). Persistent constipation and faecal incontinence occurred almost equally following any of the surgical procedures but were commoner in those undergoing surgery at a later age and in those undergoing staged operations.

The incidence of postoperative enterocolitis was higher in children who underwent surgery at an age of less than one year. Recurrent symptoms were commoner after Swenson's pull-through and in those with long segment disease but equally common among those in whom the histology of the proximal end of the resected segment was reported as irregular or aganglionic compared with those in whom it was reported as regular.

Nineteen patients underwent repeat pull through. The most common indications for redo procedures were anorectal stenosis and incomplete resection of the aganglionic colon. The Swenson's procedure was the preferred procedure in twelve patients, Soave-Boley in two and myectomy in five.

Ganglion cells are necessary for the relaxation of the bowel so that proximal contents are accepted and passed on by the process of peristalsis. The absence of transmural ganglia and of the non-adrenergic inhibitory fibres interferes with the normal relaxation mechanism of peristalsis and with internal sphincter relaxation. This in turn interferes with the normal propulsive peristalsis. Spasm, lack of propulsive peristalsis or both could account for the functional obstruction but lack of peristalsis is the most consistent cause of obstruction and pressure studies of the anal sphincter mechanism in patients with Hirschsprung's disease have disclosed that rather than distention of the rectum causing reflex internal sphincter relaxation contraction and increased internal sphincter pressure results [3].

Hirschsprung's disease results from arrested caudal migration of neuroblasts in the alimentary tract. As such no skip lesion exists with the aganglionic portion always located distally. The length of the segment varies hence the various types of megacolon [2,4].

a) Rectosigmoid (classic variation) - occurring in 70% - 85% of the cases involves the rectum and the sigmoid colon.

b) Long segment aganglionosis - occurring in 10% of the cases may extend to any level between the hepatic flexure and the descending colon.

c) Total colonic aganglionosis - occurring in 10% of the cases may involve a variable length of the terminal ileum.

d) Short segment aganglionosis – occurring in 5% of the cases involves the area just proximal to the internal sphincter mechanism [2, 4].

Presentation of Hirschsprung's disease starts at birth with delayed passage of meconium and subsequent constipation, and evacuation may occur after a rectal examination. When the passage of the first meconium is delayed beyond 48 hrs in a term otherwise healthy infant, aganglionic megacolon should be suspected [5]. Infants with Hirschsprung's disease, however, may follow different clinical courses ranging from complete intestinal obstruction at birth with vomiting, abdominal distention, failure to pass meconium and radiological evidence of low intestinal obstruction, to that of only mild constipation. In a certain group with initial mild symptoms of constipation there is an abrupt onset of enterocolitis with diarhoea, abdominal distention, fever and prostration [2].

The most serious complication in the neonatal period is ischaemic enterocolitits caused by ischaemic necrosis of the mucosa of the bowel above the aganglionic segment often extending to the small intestine. Intestinal pneumatosis, pericolic abscess, perforation and septiceamia commonly lead to death. It is the most usual cause of death in infants with untreated obstruction due to Hirschsprung's disease [6]. It can occur before and after definitive operation for Hirschsprung's disease and before or after colostomy fashioning. When present in the neonate it almost always recurs during the post-operative period [6].

Presumptive diagnosis of Hirschsprung's disease is usually made through the careful assessment of presenting history and the clinical findings of the patient. The diagnosis is, however, made after a series of investigations including plain abdominal radiographs, barium enema examination, suction rectal biopsy and full thickness rectal biopsy. Plain radiographs may show evidence of distended or dilated air filled colon with features of low intestinal obstruction. Barium enema examination may show an area of transition from dilated proximally obstructed bowel to the narrow distal segment. However 20% of cases will not have a

demonstrable transition zone especially in the newborn and in total colonic aganglionosis. Overall, radiologic diagnosis is 83% accurate, but it is inaccurate in total colonic aganglionosis, short segment disease and patients with a colostomy. In these groups of patients, anorectal manometry is superior [2]. Anorectal manomerty based on the anorectal inhibitory reflex is more reliable for the diagnosis of ultra-short segment disease. Overall its diagnostic reliability is 85% [2]. The definitive diagnosis of Hirschsprung's disease rests on the demonstration of the absence of trans-mural ganglion cells and of the presence of excess non-myelinated nerve trunks in the distal intestine in an adequate rectal biopsy. The accuracy of rectal biopsy in making a diagnosis is about 95% [2].

Suction biopsy using the Dobbin suction biopsy technique [7] obtains a specimen of mucosa and submucosa. The identification of nerve cells in the submucosal (Meissner) plexus is more difficult than in the myenteric plexus. Besides, the interpretation of ganglion cells rests on the identification of nerve units rather than large irregular ganglion cells and because nerve cells mature as the infant grows the presence as well as the number of ganglion cells is altered, as the infant grows older. The suction biopsy technique may therefore not be very helpful in making a diagnosis in a neonate [8]. Full thickness rectal biopsy from the posterior wall of the rectum taken 2-3 cm above the dentate line offers a more reliable definitive diagnosis [8].

Definitive surgical treatment of aganglionic megacolon involves bringing normal bowel as low in the rectum as is technically possible by resecting or bypassing the aganglionic bowel. In the recent years the management of Hirschsprung's disease has been changing from multistage treatment to one stage pull-through without a preliminary diverting colostomy.

Benefits of one stage treatment include avoidance of multiple operations, and elimination of complications associated with a colostomy, shorter duration of hospital stay and completion of treatment at an earlier age [9, 10].

The timing of the definitive procedure also has been changing from delayed pull-through in infants to early neonatal surgery. The decrease in incidence and severity of enterocolitis with early diagnosis and prompt decompression has encouraged earlier definitive operation often without preliminary colostomy [2]. The functional results are comparable with staged procedures; there are fewer complications and shorter hospital stay [11].

In Kenyatta National Hospital as in other developing countries staged procedures are still the main option [12], mainly due to the non-availability of laboratory facilities. A diverting colostomy is fashioned to decompress dilated colon, relief of obstruction and to protect the coloanal anastomosis after the definitive procedure. At the time of fashioning of the colostomy, full thickness serial biopsies are taken from the peritoneal reflection, the transition zone, and the dilated zone and at the site of colostomy. The histological appearance of the specimens aids in establishing the exact level of aganglionosis thereby ensuring that the pulled-through colon is normally innervated. Although staged procedures are the main practice, one-stage pull-through operations have also been performed, but the complications and outcomes of these have not been evaluated as yet [12].

The first successful definitive surgical management of Hirschsprung's disease was described by Swenson and Bill in 1948 [13]. Swenson's operation was adopted by other surgeons and over the years even in the most capable hands a regularly recurring pattern of complications became apparent. Their dissatisfaction with the

post-operative results led to the development of other procedures such as the Duhamel [14], Soave pull-through [15] and the Rehbein-States operation [16].

Swenson's Procedure

Involves resection of all of the aganglionic bowel, freeing the rectum by precise dissection close to the rectal wall down to the sphincteric mechanism, temporary closure of both the distal aganglionic rectum and the proximal normally innervated colon, eversion of the closed rectal stump, through the anus and a precise two layered anastomosis of the pulled through colon to the everted rectum performed perineally. The resection should leave at least 1.5cm of rectal wall anteriorly and almost none posteriorly so as actually to perform a posterior sphincterotomy [13].

The Duhamel's Procedure

Devised by Duhamel in 1956, the technique intended to obviate the extensive pelvic dissection in the Swenson's procedure confining the pelvic dissection to the posterior or presacral plane and retains the aganglionic anterior rectum to which the normal bowel is anastomosed.

The aganglionic intestine is resected down to the peritoneal reflection and the rectum is sutured closely. The proximal normal intestine is then brought through a retrorectal tunnel and through an incision in the posterior half of the circumference of the distal rectal wall, the posterior wall of the rectum above this level and the anterior wall of the pulled through colon then apposed by a crushing clamp which results in a wide anastomosis of the end of the colon to the posterior wall of the rectum [14].

Soave's operation

Introduced by Soave in 1964. It confines the entire pelvic dissection within the rectum. It involves removal of the mucosa of the distal bowel by submucosal dissection to the anus and the passage of the normally innervated colon through the remaining seromuscular tube.

The original procedure allowed a segment of the pulled through colon to protrude well beyond the skin margin for removal at a second stage two weeks later. Boley later modified the procedure to a one-stage procedure by primary anastomosis of the pull through colon to the seromuscular cuff at the dentate line [15].

Rehbein-State's operation

Involves extended anterior resection of the colon upto well proximal to mid-transverse colon and primary anastomosis of the transverse colon to the upper rectum. The rectum is retained and remains in continuity without alterations. Rehbein modified the original state's operation by resecting a greater portion of the rectum [16].

Post – operative complications

Conditions essential for successful treatment of Hirschsprung's disease are those common to any successful colonic operation and include adequate blood supply to the two anastomosed ends of intestine, lack of tension on the suture line, good haemostasis and adequate resection of the aganglionic intestine [2].

All the surgical procedures have been used successfully by their proponents and modifications of their original procedures have been tried by others, yet each has failed to attain the goal of complete reconstruction. Each has been attended by a recurring pattern of complications [4].

Post – operative intestinal obstruction from adhesions, volvulus or intussusception is common to all the operative procedures. Adhesion results from excessive intra-peritoneal scarring which can be avoided by gentle handling of tissues, avoidance of the use of dry pads and sponges, elimination of foreign bodies (glove powder, excessive length of sutures) and avoidance of gross ligatures of omentum and mesentery which produce nodules of necrotic tissue that invite adhesions [2].

Kleinhaus [17] reporting on an analysis of 1196 children with Hirschsprung's disease conducted by the Surgical Section of the American Academy of Paediatricians to evaluate the long term complications of major operative procedures for colonic aganglionosis excluding total colonic involvement, reported this to occur in 71% of patients undergoing the Duhamel procedure compared to 9% with Swenson's and 4.8% with the endorectal pull-through procedures. Swenson et al in their comprehensive review of 483 patients undergoing the Swenson's procedure reported this to occur in 2.7% of the patients [18].

Wound sepsis was observed in 6.5% of patients in the Swenson's et al report. Factors that predispose to wound sepsis include malnutrition, prolonged operation time, intra-operative hypothermia, inadequate bowel preparation, extensive tissue damage, inadequate hemostasis and presence of remote infection [2].

Anastomotic leakage occurred in 5.3% of the patients in the Klienhaus report [17] and was commonest following the Swenson's procedure occurring in 11.2% of the

patients who underwent the procedure compared with 2.4% with the Duhamel procedure, 1.1% with the original Soave and 5.8% with the Boley modification of Soave procedure [17]. Swenson et al in their series reported a 5% rate of anastomotic leaks [18].

Adequate preoperative bowel preparation, proper nutritional preparation and meticulous technique ensuring lack of tension, adequate blood supply and prevention of infection are important in preventing the development of anastomotic leaks and fistulae. Anastomotic leakage is particularly hazardous in the Swenson's procedure since the anastomosis is above the levator mechanism but below the peritoneal floor and therefore resulting abscess may not be apparent before rupture into the peritoneum. Anastomotic leakage in the endorotectal pull through results in a sleeve abscess between the pulled through colon and the seromuscular cuff of rectum. This may be difficult to diagnosis until chronic sepsis or fistula indicates the source of trouble [2].

Post – operative enterocolitis(POE) resulting in significant morbidity and mortality occurred in 8.3% of the patients following pull-through in the Kleinhaus report [17]. The incidence of POE according to many series ranges between 2% to 33% [19, 20,21, 22]. The survey in Klienhaus concluded that Swenson's procedure was followed by the highest incidence (15%) of POE and that the incidence of this complication was lower with the other operative procedures [1, 17]. In the Swenson et al review [18] enterocolitisis occurred immediately post-operatively in 79 (16.4%) and late in 100 (27%) of the patients comparing well with the Kleinhaus report [17].

The most common explanation of this complication is the presence of a persistently hypertonic anal sphincter resulting in a functional obstruction, leading to infection and inflammation of the proximal bowel, [22,23, 24, 25]. Late postoperative enterocolitis

may mimic symptoms of untreated Hirschsprung's disease [2]. To treat POE, sphincterotomy and anorectal myomectomy have been used with good, long-term results in most cases [26, 27].

Wilson-Storey et al by assessing immunological mucosal defence in patients who had enterocolitis showed that these patients had a marked deficiency in transfer of secretory IgA across the gastrointestinal mucosal cell, and thus were prone to mucosal invasion of both pathogenic and commensal organisms. The abnormalities were detected upon initial investigation (at presentation) prior to onset of enterocolitis, and persisted into later life. This observation helped to explain why enterocolitis can occur many years after definitive surgery [28].

Rectal stenosis/stricture is associated with all major procedures [2]. Several series quote this to be commonest following the Soave procedure [17,22, 29] but less frequent with the Boley modification. Other series quote this to be commonest with the Swenson's procedure [2]. Swenson et al in their review [18] reported it in 6.2% of their patients.

Constipation persisting after pull-through can be functional with spontaneous improvement on long term follow up [2]. It has been well established that the Duhamel procedure even with a modification to eliminate the rectal pouch is significantly more constipating than other as seen in several series [25, 29, 30, 31,32,33]. This is felt to be caused by enlargement and inadequate emptying of the partially aganglionic rectal pouch with subsequent stenosis, fecoloma and soiling [33].

Faecal incontinence/soiling occurred in 13.3% of the patients following the Swenson's procedure in his review [18]. This is believed to be due to extensive pelvic dissection and destruction of sensory nerves of the rectum undertaken in the

Swenson's procedure [2]. Severe perianal excoriation related to frequent bowel motions are known to be common after an endorectal pull through and felt to be caused by the small capacity of the neorectum created in the procedure [20, 29, 34]. With time however this has been shown to improve [2,18,30].

Ano-rectal achalasia may persist after pull-through and cause ineffective bowel evacuation with persistent constipation, faecal retention and frequent bouts of enterocolitis [33,35]. Anorectal achalasia is believed to occur because of inadequate division of the internal sphincter of the anorectum [36].

The problem with all the procedures for Hirschsprung's disease whether Duhamel, Swenson's or Soave is that a third of the abnormal internal sphincter that lies below the dentate line is left intact below the anastomosis [27], and majority of the children continue to have a relatively non-relaxing internal sphincter and persistent anorectal achalasia is not uncommon leading to abdominal distention constipation and frequent bouts of POE [17, 20, 25, 37, 38].

It has been reported by several investigators that achalasia of the internal sphincter and the rectum as well as failure of peristalsis in the aganglionic colonic segment are responsible for severe constipation [39,40]. Thus eliminating anorectal achalasia as well as resecting the anganglionic colon segment are crucial for treating Hirschsprung's disease [33,39].

Kasai-et al [39] first attempted intra-operative internal sphincterotomy at the time of colectomy and the procedure allowed the function of the anorectum be almost normal.

Marks [41] reported on the endorectal split sleeve pull through procedure in which a posterior median proctomyotomy was performed from above to within one centimetre of the dentate line to accomplish partial internal sphincterotomy.

Kimura et al described five post-Soave pull through patients in whom a posterior sagittal internal sphincterotomy was curative for persistent achalasia [42].

Takeshi Minayo et al modified the Soave pull-through by performing near-total sphincterotomy using a sagittal incision of the posterior portion of the rectum in the midline just distal to the dentate line. In a retrospective study of 43 patients undergoing the modified Soave pull through they noted a relatively low incidence (5%) of postoperative enterocolitis [43].

The surgical management of patients with Hirschsprung's disease at Kenyatta National Hospital has been changing over the years as in other places. These changes have been attributed to the need for using surgical procedures with fewer complications and the increasing number of specialist paediatric surgeons each with differing preferences for the definitive surgical procedure [12].

Barrak in a ten year rectospective study of 162 patients seen with Hirschsprung's disease in Kenyatta National Hospital revealed that the surgical procedures carried out were Soave (Boley modification) in 88 patients, Rehbein –State's operation in 56 patients, myectomy in 9 patients and Duhamel in 9 patients. The immediate post operative complications observed were wound infections, in 11%, anastomotic leaks, in 2.6% pelvic abscesses, in 2.6% and intestinal obstruction in 1.9% of the patients. Late complications included rectal stricture in 11% and temporary soiling in 8%. The highest incidence of postoperative complications was with Duhamel procedure and the best results were seen with the Soave –Boley procedure [12].

Serial biopsies taken at the time of colostomy are a significant tool in the histological diagnosis of Hirschsprung's disease and in establishing the exact level of aganglionosis, especially in instances where the transition zone delieneated by radiological examination is not certain.

The histological demarcation of the transition from aganglionic to normally innervated colon is not always dramatic and requires good pathological interpretation, yet this demarcation sets the limits for resection in the definitive procedure. This pathological differentiation of normal and aganglionic bowel is also necessary while examining the proximal ends of the resected bowel segments at the time of definitive surgery.

Schulter et al [44] reporting on the role of the histomorphological findings in the proximal segment of the resected bowel specimen in predicting post-operative functions reported a strong link between the two. A histologicaly regular proximal bowel segment generally predicted good post-operative bowel function. In patients with intestinal neuronal dysplasia (IND) of the proximal segment, the overall clinical result remained unchanged with a rise in constipation rate and in additional encopresis. The distinct group with aganglionosis of the proximal segment follows a complicated post-operative course with secondary bowel resection and recurrent episode of enterocolitis. However, these findings become less important whenever an extensive resection more than left hemicolectomy is required.

Total colonic involvement fortunately is a rare form of Hirschsprung's disease accounting for 5-8% of all cases. Signs and symptoms of total aganglionosis are more severe. Mortality rate has been reported at 65% although in recent years it has significantly been reduced due to the introduction of total parenteral nutrition and new surgical procedures [45, 46]. There are complications peculiar to the surgical

correction of total colonic aganglionosis. Initially the stools are predominantly liquid but over time become formed. Recurrent bouts of abdominal distension are common if the anastomosis is created too high on the posterior rectal wall, while incontinence is common if the anastomosis is too low for which a posterior approach and plication of the terminal ileum gives satisfactory results. Protracted diarrhoea due to regrowth of the septum between the small bowel and the rectum can be corrected by resection of the septum [2,4].

Short segment aganglionosis has for many years been managed by anorectal myectomy since it was described by Lynn 1966. The outcome depends on the resection extending well proximally to areas with normal ganglion cells other wise complications follow related to retained aganglionic segment or inadequate resection of the internal sphincter with features of anoretal achalasia [2,4].

Secondary Procedures

A secondary procedure is undertaken when the original procedure has failed or is attended by unrelenting complications. The common indications for secondary procedures are, incomplete resection of the aganglionic segment, anastomotic leak, anorectal stenosis/stricture, fistula, recurrent rectal septum, rectal prolapse, persisting incontinence and anorectal achalisia or failed pull-through [25, 36].

The commonly undertaken secondary procedures include:- Myectomy,rectal septum division, diverting colostomy, sphincterotomy, fistulectomy, revision of rectal prolapse and repeat pull-through [25, 36].

On repeat pull-through procedures, a more difficult pelvic dissection is encountered due to adhesions and difficulties in mobilising already shortened bowel while

avoiding tension and ensuring adequate blood supply to the pulled through bowel. There is, therefore, a high risk of damage to more pelvic structures especially the nerves to the vas deferens leading to infertility; and the sensory nerves to the rectum with more incontinence. Sphincter mechanism stenosis and stricture are also quite common following repeat pull-through [47].

Recurrent constipation and faecal incontinence as complications of a remote post-operative period take place more frequently than incontinence of faeces. Improvement and later complete recovery is common, so that early functional evaluation may be erroneous. A follow up period of less than five years is insignificant for the final evaluation of the various operative procedures [2,30]. One of the causes of constipation after radical operation for Hirschsprung's disease may be a long hypoganglionic zone of the distal portion of the colon. To solve the question about re-operation of children with Hirschsprung's disease evidence of aganglionosis in the distal bowel should be demonstrated in full thickness rectal biopsy [2]. Manometry should also be performed to rule out achalasia of the internal anal sphincter and intestinal neuronal dysplasia [42].

Although inadequate resection of the aganglionic bowel is the most common explanation of this aganglionosis after an otherwise well performed procedure, acquired aganglionosis must be borne in mind. Acquired aganglionosis is a rare but documented occurrence following pull-through for Hirschsprung's disease [48].

West et al [48] confirmed the presence of normal ganglion cells on the biopsies of the pulled-through segment at the initial operation. With recurrence of symptoms full column barium enema studies revealed a transition zone or narrow area in the rectosigmoid or descending colon and repeat full thickness rectal biopsy at 3cm

above the anal verge in the pulled-through segment confirmed the absence of ganglion cells.

Vascular compromise of the distal bowel segments at the time of initial pull-through procedure may contribute to the selective loss of ganglion cells post-operatively as neural tissues are most sensitive to hypoxia [48].

Even after a seemingly well-performed secondary procedure, symptoms may persist in some patients. In such patients a possible cause of failure of treatment is Intestinal Neuronal Dysplasia (IND) [49]. This owes credit to recent evidence that Hirschsprung's disease is only a part of a spectrum of conditions representing disorders of bowel innervation and ganglion distribution, that can co-exist and includes IND [50].

IND is a distinct clinical entity, which can be clearly proven histologicaly. Patients with IND not only have abnormalities of submucosal and myenteric plexuses but also defective innervation of the muscle and neuromuscular junction as well as the internal sphincter [50]. The frequency of IND co-existing with Hirschsprung's disease has been reported to vary between 20% to 66% [50].

Precise options for therapy have not been clearly established but it is worth noting that clinical regression and objective histological improvement have been reported [50]. Banani et al reporting on 11 out of 20 patients with recurrent symptoms who did not respond to posterior anorectal myectomy noted that all had histological signs of IND in previously pulled through colon. There was, however, satisfactory bowel movement in a 10–54 months follow up period following subtotal colectomy of the descending and transverse colon and pull-through of the right colon after clockwise derotation of the entire bowel. He felt that this procedure would be

effective when symptoms persist after conservative therapy or posterior anorectal myectomy [49].

RATIONAL OF THE STUDY

In Kenyatta National Hospital Hirschsprung's Disease has been managed for over 30 years. Every year approximately 11 patients with Hirschsprung's Disease undergo surgical management in the hospital. The results of that treatment are not uniformly successful and complications are noted in about 12 percent of these patients. Some have had long hospital readmission and others have even required repeat pull through procedures. The management of these complications poses an enormous challenge to the surgeon, the patient, the relative/parent and the health resources. This study is therefore aimed at critically evaluating the complications following the surgical treatment of Hirschsprung's Disease at Kenyatta National Hospital.

STUDY OBJECTIVES

Broad Objectives

To critically review the post surgical complications of Hirschsprung's disease and their management at Kenyatta National Hospital between January 1991 and December 2000.

Specific objectives

1. To study the types and rates of complications following surgical management of Hirschsprung's disease.

2. To determine whether the types and rates of complications are related to the type of surgical procedure performed, the length of aganglionic colon segment involved, the histological appearance of the proximal end of the resected colonic segment, the timing of the definitive procedure, and the staging of the definitive procedure.

3. To study the causes of, the types of, complications and outcomes of redo procedures.

MATERIALS AND METHODS

This is a descriptive retrospective study of patients who had a pull-through or other relevant surgical procedure for histologicaly proven Hirschsprung's disease over a period of 10 years from 1st January 1991 to 31st December 2000. Case records of all patients with histologically proven Hirschsprung's disease treated in Kenyatta National Hospital within the study period were retrieved from records department of Kenyatta National Hospital. Information was extracted from records of those who have undergone pull-through and fit in the admission criteria. The information obtained from these records including age, details of clinical examinations, radiologic, histopathological and intra-operative findings was reviewed to determine the type of aganglionosis, the nature of operative management, the complications and the outcome of treatment in these patients.

Histological findings of biopsies were classified as: -

a) aganglionic, if ganglion cells were entirely absent ;

b) regular, if ganglion cells were present and normal in size, maturation and distribution; and,

c) irregular, if ganglion cells were present but abnormal such as hypoganglionosis, immature, distorted or sparsely distributed.

Complications in this study were classified as: -

a) early : if occurring within one month of the definitive surgery.
b) late : if occurring later than one month after the definitive surgery.

A secondary procedure is any procedure performed after the initial intended definitive procedure either due to failure of the first operation or due to complications arising from it.

The outcomes of secondary procedures (re-operations) and overall management of Hirschsprung's disease were evaluated as:-

a) good, when there was significant improvement. The patients developed voluntary bowel movements without constipation, soiling or recurrence of symptoms.

b) average, when some improvement occurred after operation but the patient developed mild complications. The patients developed minimal soiling (once or twice per week in minimal amounts) giving no social problem to the patients; or constipation managed by change of diet or laxatives.

c) poor, when no improvement was noted or the patient developed severe complications: constipation requiring enemas; soiling that is constant and representing a social problem to the patient; recurrence of symptoms; or other related complications requiring surgical correction.

Constipation is defined as the incapacity to empty the rectum spontaneously (without help) every day.

Soiling is defined as the involuntary leakage of small amounts of stool, which provokes smearing of the underwear.

Eligibility criteria

Inclusion

- All patients operated on at Kenyatta National Hospital for histologically proven Hirschsprung's disease and any colostomies closed within the period of study.

- All patients must have had 6 months follow-up after closure of colostomy.

Exclusion

- All patients without adequate medical records for example missing histopathology reports.

- All patients whose colostomies were still open after pull-through or awaiting definitive surgery.

Study Limitations

a) Records of some of the patients were incomplete and with some of the clinical details missing for example physical findings on examination, intra-operative findings etc.

b) Some of the patients records could not be traced

c) Loss of patients to follow up.

d) Misinterpretation of patients' symptoms.

Ethical considerations

This was a retrospective study and the data was obtained from patients' records. The information obtained was handled in a manner to ensure privacy and protect confidentiality. The strategies for protecting confidentiality included:-

a) storing data in locked file cabinets,
b) limiting access to the research data,
c) coding data to hide identity of subjects, and
d) ensuring that the individual subjects cannot be identified when the findings are published.

The study was conducted after obtaining approval from Kenyatta National Hospital Ethical and Research committee.

The data was extracted using a predesigned proforma questionaire (appended) then entered into a computer and analysed using SPSS/pc+ for windows version 10.05. Analysis involved descriptive statistics like means, standard deviations, medians, frequency distributions and cross tabulations. For categorical data where comparisons were made between groups, Chi-square statistics or Fisher's exact probability (where applicable, in case chi-square was not valid) were used. The level of significance of 5% ($p<0.05$) was used.

RESULTS

In the study, medical records of 96 patients who underwent surgery for histologically proven Hirschsprung's disease and were followed up at Kenyatta National Hospital, during the period of study January 1991 to December 2000 were reviewed. Fourteen were females and 82 were males with a sex ratio, females to males of 1: 6 The age at diagnosis ranged from 3 days to 15 years with a mean age at diagnosis of 2.7 years and standard deviation of 2.95 years (mode =3 years and median = 1.71 years). Only four (4.2%) of the patients were diagnosed in the neonatal period and all were males. Of the other males 28 (34.1%) were diagnosed below one year of age, 40 (48.8%) between one year and five years, and only 14 (17.1%) after five years of age. All the females were diagnosed at an age of below four years with 8 (57.1%) being diagnosed in infancy. Using either, radiological, histopathological or intra-operative findings the patients were classified according to the length of aganglionic colon involved. They were also classified according to the age at the time of definitive surgery, the staging of operations, the surgical technique employed and the histological findings of the resected segment of colon.

Table 1

Classification of patients based on the length of aganglionic colon involved as seen at Kenyatta National Hospital.

Type	no. of patients	percentage
Rectosigmoid	75	78.1
Short segment	15	15.6
Long segment	6	6.3
Total	96	100.0

Rectosigmoid (classical) disease occurring in 78.1% of the patients was the commonest variant encountered.

Table 2

Classification of patients based on the surgical procedures performed in the management of Hirschsprung's disease at Kenyatta National Hospital.

Type of operation	No. of patients	Percentage
Swenson's procedure	54	56.3
Soave – Boley	39	40.6
Myectomy	3	3.1
Total	96	100

Only three types of procedures were performed, Swenson's pull-through, Soave – Boley pull-through and myectomy. The Swenson's pull-through was the most preferred procedure.

The resected segments of the colon were subjected to histological examination in 63 of the patients. The proximal ends of the resected segments were reported as normal (regular) in 44 (45.8%), irregular in 9 (9.4%) and aganglionic in 10(10.4%) of the patients. The histology of the resected segment was not available in 33 (34.4%) of the patients.

Timing of definitive procedures

a) Age at definitive surgery

None of the patients had definitive surgery in the neonatal period. Sixteen (16.7%) had surgery performed between 1 month and 1 year and 80 (83.3%) had surgery after one year of life.

b) Staging of operations

Seventeen (17.7%) patients underwent one-stage pull-through (without colostomy); 17 (17.7%) two-stage and 62 (64.6%) had a three-staged procedure. Three of the patients undergoing two-stage and three-stage procedures had colostomy fashioned at the time of definitive surgery. The other 76 had a preliminary decompressing colostomy and serial biopsies taken at the same time.

Post-surgical complications of Hirschsprung's disease observed at Kenyatta National Hospital

Table 3 and 4 show the various complications observed following the definitive surgical procedures for Hirschsprung's disease.

Early complications

Out of the 96 patients who underwent surgery, only 17 (17.7%) developed complications in the immediate post-operative period while 79 (82.3%) had no complications. The main complications observed are shown in table 3.

Table 3

Complication	**No. of patients with complication**	**Percentage**
Wound Infection	8	8.3
Enterocolitis	4	4.1
Anastomotic leak	2	2.0
Pelvic abscess	3	3.1
Intestinal Obstruction	3	3.1
Wound dehiscence	2	2.0
Peritonitis	1	1.0

Some patients had more than one complication. Wound infection was the commonest early complication

Late post-operative complications were observed in 70 (72.9%) of the patients. The complications observed are shown in table 4.

Table 4

Late complications

Complication	No. of patients with complication	Percentage
Persistent constipation	25	26.0
Incontinence	21	21.9
Rectal stenosis	20	20.8
Late POE	11	11.5
Recurrent symptoms	12	12.5
Intestinal obstruction	4	4.2
Rectal prolapse	3	3.1
Fistulae	3	3.1

Some patients had more than one complication occurring simultaneously or at different times during follow up. The main complications included persistent constipation, faecal incontinence, anorectal stenosis, post-operative enterocolitis and recurrent symptoms. Of note is the increased number of patients developing enterocolitis compared with those who developed it in the immediate post-operative period.

Colostomy related complications

Of the 76 patients who underwent a preliminary decompressing colostomy; 51 (67.1%) had no colostomy related complications. However, 20 (26.3%) developed stomal prolapse, 3(3.9%) developed stomal stenosis and 2(2.6%) developed colostomy stomal bleeding. Three patients required refashioning of the colostomies.

Table 5 (a)

Early complications observed after correction of the various levels of aganglionosis

	Length of colon involved					
	rectosigmoid		Short segment		Long segment	
Complications	**no.**	**%**	**No.**	**%**	**No.**	**%**
Wound infection (8)	7	87.5	1	12.5	0	0
Anastomotic leak (2)	2	100	0	0	0	0
Pelvic abscess (3)	3	100	0	0	0	0
Early POE (4)	3	75	1	25	0	0
Intestinal obstruction (3)	3	100	0	0	0	0
Wound dehiscence (2)	2	100	0	0	0	0
Peritonitis (1)	1	100	0	0	0	0

Most of the complications were observed after correction of rectosigmoid disease though majority of the patients had rectosigmoid disease. Due to the small numbers involved further statistical tests were invalid.

Table 5(b)

Late complications observed after correction of the various levels of aganglionosis

	Length of colon involved					
	rectosigmoid		short segment		Long segment	
Complications	**no.**	**%**	**No.**	**%**	**No.**	**%**
Ano-rectal stenosis (20)	18	90	1	5.6	1	5.6
Persistent constipation (25)	20	80	4	16	1	4
Incontinence (21)	13	61.9	7	33.3	1	4.8
Late POE (11)	9	81.8	2	18.2	0	0
Recurrent symptoms (12)	7	58.3	2	16.7	3	25.0
Intestinal obstruction (4)	3	75	1	25	0	0
Rectal prolapse (3)	3	100	0	0	0	0
Fistulae (3)	3	100	0	0	0	0

Majority of the complications occurred among those patients with rectosigmoid disease. The statistical significance of this was however difficult to test because both Chi-square and Fisher's exact tests were invalid.

Twenty-five percent (25%) of recurrent symptoms were in the patients with long segment disease. However, this represented a significantly large number of patients (50%) among those with long segment disease and this was found to be statistically significant (p value = 0.01).

Table 6 (a)

Early complications observed against the major operative procedures

	Surgical technique					
	Swenson's		Soave-Boley		Myectomy	
Complications	**no.**	**%**	**No.**	**%**	**No.**	**%**
Wound infection (8)	3	37.5	5	62.5	0	0
Anastomotic leak (2)	0	0	2	100	0	0
Pelvic abscess (3)	0	0	3	100	0	0
Early POE (4)	3	75	1	25	0	0
Intestinal obstruction (3)	2	66.7	1	33.3	0	0
Wound dehiscence (2)	1	50	1	50	0	0
Peritonitis (1)	1	100	0	0	0	0

The Soave-Boley procedure had a higher complication rate, compared with other procedures. Since there were no complications noted among the myectomy group, comparison was done between the Swenson's and the Soave-Boley groups.The Soave-Boley group had a higher complication rate compered with the Swenson's group but the differences for individual complications were statistically insignificant (Fisher's exact probability, p value >0.07 in all).

Table 6 (b)

Late complications observed against the major operative procedures

	Surgical technique					
	Swenson's		Soave-Boley		Myectomy	
Complications	**no.**	**%**	**No.**	**%**	**No.**	**%**
Rectal stenosis (20)	9	45	11	55	0	0
Persistent constipation (25)	12	48	12	48	1	4
Incontinence (21)	11	52.4	10	47.6	0	0
Late POE (11)	6	54.5	5	45.5	0	0
Recurrent symptoms (12)	8	66.7	3	25	1	8.3
Intestinal obstruction (4)	3	75	1	25	0	0
Rectal prolapse (3)	0	0	3	100	0	0
Fistulae (3)	0	0	3	100	0	0

Persistent constipation and faecal incontinence were almost equaly as frequent in the Swenson's and the Soave-Boley groups.

Recurrent symptoms were however commoner in the Swenson's group, but the statistical significance of this could not be evaluated because the Chi-square test was invalid.

Rectal stenosis was commoner after the Soave-Boley procedure (55%) compared with the Swenson's procedure (45%). However, this was statistically insignificant (Fisher's exact probability, p value = 0.18).

Table 7 (a)

Early complications observed with different staging of the correction of Hirschsprung's disease

	Staging of operations					
	One-stage		Two-stage		Three-stage	
Complications	**no.**	**%**	**No.**	**%**	**No.**	**%**
Wound infection (8)	1	12.5	1	12.5	6	75
Anastomotic leak (2)	0	0	0	0	2	100
Pelvic abscess (3)	1	33.3	0	0	2	66.7
Early POE (4)	1	25	0	0	3	75
Intestinal obstruction (3)	1	33.3	1	33.3	1	33.3
Wound dehiscence (2)	0	0	0	0	2	100
Peritonitis (1)	0	0	0	0	1	100

Intestinal obstruction occurred as frequently in all the three groups of patients but there was a higher incidence of the other complications in those who underwent three-stage procedures. Tests for statistical significance were invalid

Table 7(b)

Late complications observed with different stagings of the correction of Hirschsprung's dissease

	Staging of operations					
	One-stage		Two-stage		Three-stage	
Complications	**no.**	**%**	**no.**	**%**	**No.**	**%**
Rectal stenosis (20)	4	20	0	0	16	80
Persistent constipation (25)	5	20	6	24	14	56
Incontinence (21)	4	19.0	5	23.8	12	57.1
Late POE (11)	4	36.4	2	18.2	5	45.5
Recurrent symptoms (12)	1	8.3	3	25	8	66.7
Intestinal obstruction (4)	1	25	0	0	3	75
Rectal prolapse (3)	2	66.7	0	0	1	33.3
Fistulae (3)	0	0	0	0	3	100

Persistent constipation and incontinence were commoner after three-stage procedures compared with one-stage and two-stage procedures but this was statistically insignificant (p values = 0.53 and 0.66 respectively).

Late post-operative enterocolitis occurred almost as frequently in the one-stage and the three-stage groups but less frequently in the two-stage group but this was statistically insignificant (p value = 0.21).

Intestinal obstruction occurred more frequently after three-stage (75%) compared with one-stage procedures (25%) though statistically insignificant (p value = 0.6).

Table 8(a)

Early complications observed against the histological reports of the proximal ends of resected segments of colon

	Histological reports							
	Regular		Irregular		Aganglionic		Not available	
Complication	no.	%	no.	%	no.	%	no.	%
Wound infection (8)	6	75	0	0	0	0	2	25
Anastomotic leak (2)	1	50	0	0	0	0	1	50
Pelvic abscess (3)	1	33.3	0	0	0	0	2	66.7
Early POE (4)	1	25	0	0	0	0	3	75
Intestinal obstruction (3)	1	33.3	0	0	1	33.3	1	33.3
Wound dehiscence (2)	2	100	0	0	0	0	0	0
Peritonitis (1)	1	100	0	0	0	0	0	0

The histological reports of the proximal ends of resected segments bore no relevance to the development of early complications and these occurred almost randomly in those with regular histology and those in whom the reports were not available.

Table 8 (b)

Late complications observed against the histological reports of the proximal ends of resected segments of colon

	Histological reports							
	Regular		Irregular		Aganglionic		Not available	
Complication	**No.**	**%**	**No.**	**%**	**No.**	**%**	**No.**	**%**
Rectal stenosis (20)	12	60	1	5	2	10	5	25
Persistent constipation (25)	9	36	1	4	1	4	14	56
Incontinence (21)	11	52.4	3	14.3	1	4.8	6	28.6
Late POE (11)	4	36.4	2	18.2	1	9.1	4	36.4
Recurrent symptoms (12)	4	33.3	2	16.7	2	16.7	4	33.3
Intestinal obstruction (4)	3	75	0	0	0	0	1	25
Rectal prolapse (3)	0	0	0	0	1	33.3	2	66.7
Fistulae (3)	1	33.3	1	33.3	1	33.3	0	0

Persistent constipation occurred more frequently in those in whom the histological reports were not available (56 %) compared with those in whom the reports were available (44%). This was however not statistically significant (Fisher's exact probability, p value = 0.07).

Recurrent symptoms developed equally in those with regular histological reports and those in whom the reports were not available (33.3%); and even in those in whom it was reported as irregular and aganglionic combined. Though statistically insignificant (p value = 0.4), it is concerning to note that in such a large proportion

of patients developing complications (33.3%), histological reports of proximal ends of the resected segments were not available.

Post operative enterocolitis occurred as frequently in those with regular reports and in those in whom the reports were not available. This is understandable bearing in mind the multiplicity of factors involved in the pathogenesis of enterocolitis of Hirschsprung's disease.

Table 9 (a)

Early complications observed against age at definitive surgery

	Age at definitive surgery			
	1 month - 1 year		More than 1 year	
Complications	**No.**	**%**	**No.**	**%**
Wound infection (8)	0	0	8	100
Anastomotic leak (2)	0	0	2	100
Pelvic abscess (3)	0	0	3	100
Early POE (4)	0	0	4	100
Intestinal obstruction (3)	1	33.3	2	66.7
Wound dehiscence (2)	0	0	2	100
Peritonitis (1)	0	0	1	100

Most of the complications occurred in those operated on after 1 year of life though majority (83.3%) of the patients were operated after 1 year of life.

Table 9 (b)

Late complications observed against age at definitive surgery

	Age at definitive surgery			
	1 month - 1 year		More than 1 year	
Complications	**No.**	**%**	**No.**	**%**
Rectal stenosis (20)	2	10	18	90
Persistent constipation (25)	5	20	20	80
Incontinence (21)	2	9.5	19	90.5
Late POE (11)	4	36.4	7	63.6
Recurrent symptoms (12)	2	16.7	10	83.3
Intestinal obstruction (4)	0	0	4	100
Rectal prolapse (3)	2	66.7	1	33.3
Fistulae (3)	0	0	3	100

Persistent constipation and incontinence occurred more frequently in those who underwent definitive surgery at an age of more than one year (80% and 90.5% respectively) compared with those who underwent definitive surgery at an earlier age (20% and 19% respectively), though these were statistically insignificant (p values = 0.4 and 0.2 respectively).

An appreciable percentage of late post-operative enterocolitis developed in those who had surgery performed at an age of more than one year of life (63.6%) compared with those who had surgery at an earlier age (36.4%) but this was statistically insignificant (p value = 0.08)

Secondary procedures/ re-operations

Of the 96 patients who underwent surgery for the various forms of Hirschsprung's disease, 35 (36.5%) developed complications that required a secondary surgical procedure to correct. The causes of these secondary procedures/re-operations are shown in table 10.

Table 10

Causes of re-operations

Indications	**no. of patients**	**Percentage**
Anorectal stenosis	18	48.6
Incomplete resection	7	18.9
Recurrent symptoms	3	8.1
Intestinal obstruction	3	8.1
Rectal prolapse	2	5.4
Fistulae	3	8.1
Pelvic abscess	1	2.7
Total	**37**	**100**

Table 11

The main operations performed during re-operation

Procedures	No. of patients	Percentage
Repeat pull through	14	37.8
Sphincterotomy	5	13.5
Manual anal dilatation	9	24.3
Laparotomy	3	8.1
Revision of rectal prolapse	2	5.4
Fistulectomy	1	2.7
Incision and drainage	1	2.7
End colostomy	2	5.4
Total	**37**	**100**

Two patients required more than one operation.

i) Laparotomy for intestinal obstruction and sphincterectomy for incomplete resection of the aganglionic colon confirmed as aganglionic by repeat full thickness rectal biopsy.

ii) Fistulectomy and repeat pull-through for incomplete resection of the aganglionic colon confirmed by repeat full thickness rectal biopsy

The outcome of re-operations was good in 17 (48.6), average in 12 (34.3%) and poor in 6 (17.1%) patients.

Table 12

Causes of re-do pull-through and the procedures performed.

	Surgical technique		
Indication	**Swenson's**	**Soave-Boley**	**Myectomy**
Incomplete resection	5	0	2
Anorectal stensosis	4	2	3
Recurrent symptoms	3	0	0
Total	12	2	5

Out of the 35 patients who underwent re-operation, 19 (54.3%) required repeat definitive surgery either as a pull-through procedure or myectomy (sphicterotomy) Out of these 19 patients, 7 patients had incomplete resection of the aganglionic colon, 2 of which were reported as irregular and 5 as aganglionic on repeat rectal biopsy.

Persistent constipation, incontinence and enterocolitis were the main complications noted. These however improved after long-term follow-up.

Complications of re-do procedures

Nineteen patients underwent re-do procedures, 14 of whom developed complications. The main complications observed are shown on table 13 (a & b).

Table 13 (a)

Early complications observed after re-operation

Complications	No. of patients	Percentage
Wound infection	5	26.3
Early POE	1	5.3
Anastomotic leak	1	5.3
Pelvic abscess	1	5.3

Table 13 (b)

Late complications observed after re-operation

Complications	No. of patients	Percentage
Incontinence	7	36.8
Persistent constipation	3	15.8
Fistulae-in-ano	4	21.1
Late POE	4	21.1
Rectal prolapse	1	5.3
Intestinal obstruction	1	5.3
Recurrent symptoms	1	5.3

Follow - up

The age at last follow-up ranged between three months and eighteen years with a mean of 6.03 years (standard deviation = 3.66, mode = 4.5 years, median = 5.13 years). The duration of follow-up ranged between eight months and twelve years with a mean follow-up period of 3.24 years (standard deviation = 2.02, mode = 2.0 years, median = 5.12 years.

Table 14

Duration of follow-up	Percentage of patients
Up to 1 year	11.6
> 1 year	88.4
> 2 years	63.2
> 3 years	54.2
> 4 years	26.3
> 5 years	13.7

Final outcome of management of Hirschsprung's disease at Kenyatta National Hospital

Table 15

Outcome	Number of patients	Percentage
Good	70	72.92
Average	21	21.87
Poor	5	5.21
Total	96	100

Among those with average outcomes the main complication was either residual faecal incontinence or persistent constipation that were expected to improve on continued follow-up. All those with poor outcomes needed another re-do procedure.

DISCUSSION

Given the complexities of etiology and genetics, pathology and pathophysiology of Hirschsprung's disease it is not surprising that what appears to be well-defined and anatomicaly based surgical therapy still leads to inconsistent and poorly predictable outcomes. Complications after treatment for Hirschsprung's disease are not uncommon. Various factors influencing these outcomes can be found in the literature including age of the child at the time of treatment, extent of the disease, associated anomalies (eg Trisomy 21), the type of surgical procedure performed with their associated complications and the age of the child at follow-up.

It is generally felt that re-establishment of continuity of the gastrointestinal tract at an earlier age in childhood results in children having better control of defaecation [30]. Theoretically, early sensation of passage of feaces through the anal canal should allow early reestablishment of the anorectal reflex and other neural curcuits required for optimal control of defaecation [30].

Most of the patients (83.3%) had definitive surgery after one year of life and there was no patient who underwent definitive surgery in the neonatal period. Overall there was no significant difference in the complication rates among the age groups. However, there was a higher rate of persistent constipation and incontinence in the patients undergoing surgery at an age of more than one year. This is however expected to improve with time on follow-up, [2,18,30].

Most (78.1%) of the patients had the rectosigmoid variant of Hirschsprung's disease while 15.6% had short segment and 6.3% had long segment aganglionosis.

None of the patients had total colonic involvement. The incidence of complications was almost equal in all groups of patients. However almost half of the patients with long segment disease had recurrent symptoms after seemingly thorough surgical correction. The proximal end of the resected segment of colon was reported as aganglionic in one but normal in the other two. At repeat biopsy the patient with an earlier on aganglionic pulled through colon remained aganglionic while the other two turned to be irregular. It was difficult to ascertain whether this was due to pathological misreporting or truly cases of acquired aganglionosis, which though rare has been documented [48].

The most common surgical procedure used was the Swenson's pull-through (56.3%). In an earlier report [11], the commonest procedure employed in Kenyatta National Hospital was the Soave-Boley endorectal pull-through and no patient underwent the Swenson procedure. This was attributed to a change in the preference of procedures used earlier due to associated complications and surgeons' preferences. In this study there was no significant difference in the overall complications with the different surgical procedures. There was however a higher incidence of pelvic abscesses and anorectal stenosis following the Soave-Boley procedures than in other procedures.Recurrent symptoms were on the other hand more frequent following the Swenson's procedure while persistent constipation and incontinence occurred almost as frequently following either the Swenson's or the Soave-Boley procedures. All the patients who developed rectal prolapse had a Soave-Boley procedure.

In staged procedures colostomy related problems were frequently seen. Langer et al. [51] described stoma related complications in 26% of their two stage procedures when comparing the outcomes to a series of primary pull-throughs. In this study

32.9% of the patients undergoing staged procedures had colostomy related complications mainly stomal prolapse in 26.3% and stomal stenosis (3.9%). Indeed avoidance of an initial colostomy is one argument used to support primary pull-through procedures. Persistent constipation, incontinence post-operative intestinal obstruction developed more frequently following three-stage procedures.

Successful treatment of Hirschsprung's disease is largely determined by adequate resection of the aganglionic segment of colon as confirmed by the demonstration of normal ganglion cells at the proximal end of the resected bowel. In 44 (45.8%) of the patients the pull-through colon had normal ganglion cells. In 9 (9.4%) the ganglion cells were present but reported as either immature, sparsely distributed or distorted and in another 10 (10.4%) the pull-through end was reported as aganglionic. In another 33% of the patients the histology reports were not available.

Late post-operative enterocolitis occurred as frequently in those in whom the histological reports of proximal ends of resected segments were regular (36.4%) and in those in whom the reports were not available (36.4%).
Recurrent symptoms also occurred equally as frequently in those with regular histological reports and those in whom the reports were not available (33.3%). Though not statistically significant, this finding may seem to explain the need for getting histology on all the resected segments.
Post-operative enterocolitis occurred even in those in whom the pulled through colon had normal ganglion cells. Infact it was observed that the incidence of late post-operative enterocolitis was higher than that of early enterocolitis among the patients with normal pulled through colon.This could be explained by the distention of the the neorectum that occurs with time after definitive surgery

resulting in faecal stasis and invasion by pathogenic organisms especially if there is achalasia of the internal anal sphincter [22,23,24,25]. Furthermore, there are other factors to explain the development of enterocolitis in Hirschsprung's disease including the relative immunodefficiency among those who develop enterocolitis [28].

Though there was a no significant difference in the complication rates in the different categories of patients based on the histology of the proximal ends of the resected segments, its worth noting that 73% of the patients who developed complications were in the category with either an abnormal or absent histology.

It is interesting to note that among the ten patients in whom the pull-through colon was reported as aganglioninc, only a half of them developed complications with some patients developing more than one complication. Two developed recurrent symptoms and repeat rectal biopsy confirmed incomplete resection of the aganglionic segment and after repeat pull-through they did well. Another two had repeat pull-through due to persistent constipation attributed to anorectal stenosis and repeat biopsy for histology not performed. It would be expected that a repeat rectal biopsy be performed in all those who had been reported as having aganglionic pull-through ends to remove uncertainty about the credibility of the histology reports but this was not the case in this study. Repeat biopsy was performed in only 2 patients after they developed recurrent symptoms and it actually did confirm incomplete resection.

Anastomotic leakage occurred in 2(2%) of the patients compared with 5.3% in the Kleinhaus report [17]. Both had rectosigmoid disease and underwent 3-stage Soave-

Boley endorectal pull-through. Soave reported it in 1.1% and 5.8% in the Boley modification [17].

Though commonest in the Swenson's procedure [17] it was not reported in any of the patients who underwent Swenson's pull-through in this study inspite of this being the preferred procedure. In one patient anastomotic leak led to the development of pelvic abscess and later fistulae-in-ano accompanied by rectal prolapse and the patient was managed with an end colostomy. In the other patient the leak closed on conservative management but the patient later developed anorectal stenosis that improved after serial manual anal dilatations.

The diagnosis of post operative enterocolitis at Kenyatta National Hospital was mainly clinical defined as the occurrence of a clinical syndrome consisting of diarhoea, fever, abdominal distention, cramping abdominal pain and lethergy. The incidence of POE according to many reports ranges between 2% to 33% [19,20,21,22]. Kleinhaus [17] reported that Swenson's procedure was followed by a higher incidence (15%) of POE and that the incidence was lower in the other procedures. In this study early POE occurred in 4(4.1%) and lately in 11(11.5%) of the patients. Early POE was commoner in the Swenson's pull-through group occurring in 75% compared with 25% in the Soave-Boley group.Late POE was also commoner in the Swenson's group (54.5%) compared with Soave-Boley group (45.5%). Overall there was no statistically significant difference noted between the patients undergoing the different procedures.

There was no relationship between the development of POE and the staging of surgery. It was however commoner among those undergoing pull-through at an age of more than one year at the time of surgery. It was also commoner in rectosigmoid disease and if the pulled-through colon had abnormal or no ganglion

cells. Overall however, the difference was either not statistically significant or could not evaluated.

Though there are changing trends from delayed pull-through in infants to early primary neonatal surgery the relative immuno-incompetence of the neonate and the younger children may result in a more profound septic state compared to the older child [30]. This should raise some element of concern with primary surgical correction in the neonatal period. Almost all the patients who developed enterocolitis responded to medical management without need for surgical intervention.

Twelve patients developed recurrent symptoms. Though commoner in patients who underwent the Swenson's procedure (66.7%), it was not related to the age at definitive surgery or the staging of the operations. These occurred more frequently in those in whom the histological reports were not available compared with the group with regular reports. This could be explained by the presence of a retained segment of aganglionic colon. Out of the twelve patients only seven had histologicaly demonstrable retained aganglionic segment of colon. Two had anorectal stenosis and three were due to unexplained reasons and were assumed to be due to anorectal achalasia. However, all of these had a repeat pull-through.

Persistent constipation even after a seemingly successful definitive procedure can be functional with spontaneous improvement on long term follow-up [2]. It occurred in 25 (26%) of the patients. Upon digital rectal examination 20 of these had anorectal stenosis. The other five had functional constipation and on long-term follow-up improved without need for surgical invention. Majority did well on

laxatives. Infact ano-rectal stenosis may not necessary have been the cause for the constipation given that in some it persisted even after correction of the stenosis.

It occurred as frequently (48%) following both the Soave-Boley and Swenson's procedures. Conversely it was commoner in children undergoing surgery after one year of age (80%) compared with the younger child (20%) and in three-staged procedures.

Ano-rectal stenosis occurred in 20 (20.8%) of the patients. It was commonest after the Soave-Boley pull-through (55%) compared with Swenson's pull-through (45%). Swenson [18] reported it only 6.2% of his patients. Several series quote this to be commonest following the Soave procedure [17,22,29], while others quote this to be commonest after Swenson's pull-through [2]. Eighteen of the patients needed a secondary procedure because of either persistent constipation or recurrent symptoms. Nine were put through a programme of serial manual anal dilatations and improved. Six underwent repeat pull-through and three underwent myectomy. The introduction of a programme of dilatations after wound healing may be beneficial to avoid the development of a stenosing ring at the anastomotic line.

Faecal incontinence occurred in 21 (21.9%) of the patients. Swenson in his series reported it in 13.3% of his patients. In this study the incidence of faecal incontinence postoperatively was almost equal in those undergoing either Swenson's or Soave –Boley procedures. Destruction of the sensory nerves in the rectum due to the extensive pelvic dissection undertaken in the Swenson's procedure [2] and the small capacity of neorectum created in the endorectal pull-through procedure [20 29 34] are possible explanations of incontinence.Though

insignificant it was common after staged pull-through and in those undergoing surgery after one year of life, may be due to re-establishment of gastro intestinal continuity later. Assessed objectively majority of the patients will achieve normal bowel function as they reach adulthood. In this study majority improved on follow-up.

Post operative intestinal obstruction from adhesions, intususception or volvulus is common to all the operative procedures [3]. In this study it occurred in the early post operative period in three patients, and in the late post-operative period in four patients.It occurred more commonly following three-stage procedures. Of these, only three required laparatomy, the others being managed conservatively with success. At laparotomy one had intususception and the other two had adhesions.

Pelvic abscess followed the correction of rectosigmoid disease in 3 patients. In one it was a consequence of anastomotic leak following Soave-Boley pull-through. The patient later developed fistulae-in-ano and rectal prolapse for which an end colostomy was fashioned. He benefited later from a posterior sagittal anorectoplasty (PSARP) and pull-through. The other two patients who developed pelvic abscess after Swenson's and Soave-Boley procedures respectively did well on intravenous antibiotics with incision and drainage performed on only one of them.

Three patients developed fistulae-in-ano all after a Soave-Boley pull-through. Fistulectomy was performed in one. The other two had end colostomies fashioned. One of the two later developed sepsis and intestinal obstruction and inspite of laparoptomy died of sepsis. The other had PSARP and pull-through later and did well at follow-up though still incontinent of stool at the last follow-up visit.

Three patients developed rectal prolapse after Soave-Boley procedure. This is not a commonly described problem in the literature.

Secondary Procedures

Thirty-five patients (36.5%) required surgery for the management of complications arising after pull-through. Common indications for secondary procedures were anorectal stenosis (48.6), incomplete resection of the aganglionic colon (21.6%), intestinal obstruction (8.1%) and recurrent symptoms (5.4%)

Persistent constipation and faecal retention quite common as in other studies was noted in 25 (26%) of the patients. Upon digital rectal examination, anorectal stenosis was observed in 20 of these. Of these 20 patients, 18 had significant stenosis requiring surgical intervention. Manual anal dilatation was successful in 9, while 6 underwent repeat pull through procedure and 3 myectomy.

Twelve patients had recurrent symptoms of Hirschsprung's disease. In 2 of these anorectal stenosis was detected on digital rectal examination and after myectomy had good outcomes. Repeat rectal biopsy was performed in only 7 of these and all were noted to have an element of incomplete resection of the aganglionic colon. The retained segment in some was quite short as shown by supplementary barium enema examination. Two of these had favourable outcomes following myectomy while 5 underwent a repeat pull-through procedure.

Another nine patients underwent a repeat pull-through procedure, six of them due to anorectal stenosis and three due to recurrent symptoms.In those who developed recurrent symptoms, digital rectal examination did not reveal anorectal stenosis. A

repeat full thickness rectal biopsy was not performed and yet a repeat pull-through was performed. The recurrence of symptoms could have been explained as due to achalasia of the internal anal sphincter. These would have benefited more from an internal sphicterotomy as described by Kasai-et-al [39] without the need for a repeat pull-through.

Overall nineteen redo procedures were performed but these were justified by demonstration of aganglionosis in full thickness rectal biopsy in only seven. In two of these redo procedures the outcome was poor.

The Swenson's procedure was the preferred procedure in 12, Soave in 2 and myectomy in 5 patients. Technically the Swenson's procedure is regarded as easier as it avoids the difficult submucosal disection in already scarred tissues expected in a Soave procedure.

The outcome of re-do operations was good in 5 (26.3%) of the 19 patients. Re-do operations in addition to being technically more difficult are associated with higher complication rates. The main complication noted was faecal incontinence though in a majority of the patients in resolved spontaneously.

Five patients had poor outcomes requiring additional surgical procedures to correct these. One patient had myectomy after repeat full thickness rectal biopsy was reported as aganglionic. He developed recurrent symptoms with persistent constipation and recurrent bouts of enterocolitis. Sphincteromyotomy solved the problem. The other four patients had repeat pull-through procedures. Unfortunately none had a repeat full thickness rectal biopsy prior to the repeat pull-through. All had serious complications ranging from persistent constipation (2), ano-rectal stenosis (1) fistulae-in-ano (3), and gangrene of the pulled through colon

(1). They all had repeat pull-through procedures, either as Swenson's or Soave-Boley procedures. The outcomes of these additional pull-throughs were not evaluated.

At the end of follow-up, 26(27.08%) of the patients had a poor or average outcome the main complications being persistent constipation and soiling. The outcome was however regarded as good in 70(72.92%) of the patients. Persistent constipation and soiling however improve on long term follow-up but this requires long periods of follow-up [2,30]. In the study though, only 13.7% of the patients were followed up for more than 5 years with a mean follow-up period of 3.2years.

CONCLUSIONS

The surgical management of Hirschsprung's disease at Kenyatta National Hospital as in many other centres is associated with a recurring pattern of complications and at almost comparable rates. The types and rates of these complications cannot however be associated with the length of colon involved, the surgical technique employed in the correction of the disease, the age at definitive surgery or the staging of the operations. Since patients are diagnosed at different ages in Kenyatta National Hospital and there is no procedure that can be claimed to be superior to the other in terms of risks of developing any of the many different post-operative complications the choice of procedure to employ or the staging of operations, regardless of the age of the patient should be at the discretion of the individual paediatric surgeon.

Following a seemingly well performed operation a significant number of specimens of the resected colon were not subjected to histological examination to confirm that the pulled through colon is normal and therefore that there is adequate resection. This creates confusion when complications arise and the decision to re-operate is then not fully justified. Repeat full thickness rectal biopsy would then be of help coupled with other relevant clinical and radiological observations to make a rational decision to re-operate. The rate of repeat rectal biopsy was notable low. There should be a policy to make sure that all properly tagged specimens of resected gut are subjected to histological examination and the report availed before discharge of the child from hospital. Preferably the specimen should be examined by a pathologist inclined towards paediatric gastroenterology to avoid certain confusing reports such as distorted ganglion cells without a clear conclusion.

Some of the indications for repeat pull-through are not justified. Anorectal stenosis was a common indication for repeat pull-through. Sphincteromyotomy and/or incision of the stenosis ring may have sufficed especially considering_that most of the patients presented with persistent constipation that could as well be functional. It should be defined clearly what patients require repeat pull-through especially noting the complications that accompany repeat pull-throughs.

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APPENDIX I

Proforma questionaire

Patient's name _____

Ip. No: _____

Serial no: _____

1. Sex a) male ( )
   b) female ( )

2. Age at diagnosis _____

3. Diagnosis confirmed by
   A) plain radiographs ( )
   B) ba enema ( )
   C) suction rectal biopsy ( )
   D) full thickness rectal biopsy ( )
   E) anorectal manometry ( )

4. Colostomy fashioned & serial biopsy taken
   A) yes ( )
   B) no ( )

5. Length of aganglionic colon involved
   A) rectosigmoid (classic) ( )
   B) long segment ( )
   C) total colonic ( )
   D) short segment ( )

6. Age at definitive surgery
   A) birth to one month ( )
   B) 1 month – 1 year ( )
   C) > 1 year ( )

7. Type of operation
   A) swenson's procedure ( )
   B) soave – boley procedure ( )
   C) myectomy ( )
   D) duhamel procedure ( )
   E) martin's procedure ( )
   F) rehbein-state's operation ( )

8. Staging of operation
   A) one - stage ( )
   B) two-stage ( )
   C) three-stage ( )

9. Histological report of proximal end of resected segment
   A) regular ( )
   B) irregular ( )
   C) aganglionic ( )

10. Colostomy related complications

   A) prolapse ( )
   B) retraction ( )
   C) stenosis ( )
   D) bleeding ( )
   E) parastomal hernias ( )
   F) others specify _____

11. Early complications

   A) wound infections ( )
   B) wound dehiscence ( )
   C) anastomotic leak ( )
   D) pelvic abcess ( )
   e) early enterocolitis ( )
   F) intestinal obstruction ( )

12. Late complications

A) late post-operative enterocolitis ( )
B) persistent constipation ( )
C) faecal incontinence/soiling ( )
D) rectal prolapse ( )
E) recurent rectal septum ( )
F) fistulae ( )
g) rectal stenosis/stricture ( )
H) recurrent symptoms ( )
I) intestinal obstruction ( )

13. Secondary procedures needed

A) yes ( )
B) no ( )

14. Histology report of repeat biopsy if done

A) regular ( )
B) irregular ( )
C) aganglionic ( )

15. Indications for secondary procedure

A) ano-rectal stenosis ( )
B) persistent constipation ( )
C) retained aganglionic sement ( )
D) recurrent symptoms ( )
E) fecal incontinence ( )
F) fistulae ( )
G) rectal prolapse ( )
H) recurrent rectal septum ( )
I) intestinal obstruction ( )

16. Type of procedure performed

A) laparatomy for intertinal obstruction ( )
B) myectomy ( )
C) division of rectal septum ( )
D) revision of rectal prolapse ( )
E) fistulectomy ( )
F) repeat pull-through procedure ( )
G) others (specify) _____

17. Type of pull-through performed in seconday procedure

A) swenson's procedure ( )
B) soave-boley procedure ( )
C) Myectomy/sphincterotomy ( )
D) Duhamel procedure ( )
D) rehbein-state's operation ( )
E) others (specify) _____

18. Early complications

A) wound infection ( )
B) wound dehiscence ( )
C) anastomotic leak ( )
C) pelvic abscess ( )
D) early enterocolitis ( )
E) intestinal obstruction ( )
F) others (specify) _____

19. Late complications

A) late poe ( )
B) persistent constipation ( )
C) faecal incontinence/soiling ( )
D) rectal prolapse ( )
E) recurrent rectal septum ( )
F) fistulae ( )
G) rectal stenosis ( )
H) recurrent symptoms ( )
I) intestinal obstruction ( )
J) others (specify) _____

20. Outcome of secondary procedure

A) good ( )
B) average ( )
C) poor ( )

21. Re-operation needed

A) yes ( )
B) no ( )
C) if yes (specify) _____

22. Age at last follow-up _____

23. Duration of follow-up _____

24. Final outcome of management

A) good ( )
B) average ( )
C) poor ( )

APPENDIX II

Approval by Kenyatta National Hospital Ethical and Research Committee

Attached below is the letter of approval of the research "POST SURGICAL COMPLICATIONS OF HIRSCHSPRUNG'S DISEASE AND THEIR MANAGEMENT AT KENYATTA NATIONAL HOSPITAL:A TEN YEAR RETROSPECTIVE STUDY'' from the Kenyatta National Hospital Ethical and Research Committee.

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16 May 2002

Dr. F. Njoroge Kuria
Dept. of Surgery
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Dear Dr. Kuria,

RESEARCH PROPOSAL "COMPLICATIONS FOLLOWING SURGICAL MANAGEMENT OF HIRSCHSPRUNGS DISEASE IN KENYATTA NATIONAL HOSPITAL: A TEN YEAR RETROSPECTIVE STUDY" (P13/1/2002)

This is to inform you that the Kenyatta National Hospital Ethical and Research Committee has reviewed and **approved** the revised version of your above cited research proposal.

On behalf of the Committee I wish you fruitful research and look forward to receiving a summary of the research findings upon completion of the study.

This information will form part of data base that will be consulted in future when processing related research study so as to minimize chances of study duplication.

Thank you.

Yours faithfully,

PROF. A.N. GUANTAI
SECRETARY, KNH-ERC

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