A REVIEW OF THE MANAGEMENT OF HIRSCHSPRUNG'S DISEASE AT THE KENYATTA NATIONAL HOSPITAL (JANUARY 1991 - DECEMBER 2000)

BY

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DECLARATION

This dissertation is my original work and has not been presented for a degree in any other university or for publication in any journal.

Signed ..... Date 27-08-2002

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This dissertation has been submitted with my approval as a university supervisor.

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DEDICATION

This work is dedicated to my wife Nakera and my children Sieunda, Agutu and Obiero

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SUMMARY

A retrospective study covering 10 years (January 1991 to December 2000) was carried out on the management of Hirschsprung's disease at Kenyatta National Hospital. Ninety-three cases certified the inclusion criteria as underlined below.

There were 75 males (80.6%) and 18 females (19.4%). The age at histological diagnosis ranged between 14 days to 13 years with the majority (50.4%) being in the age range of 1 to 5 years.

The majority of patients were operated in age range of 1 to 5 years (59.1%). No patient below the age of one month was operated. The most common presenting complaints were abdominal distension, constipation and failure to pass meconium within 48 hours of birth. All patients had a histological diagnosis of the disease. The other common diagnostic methods used were laparatomy and serial biopsy (75.3%) and barium enema radiography (69.9%). Most patients (93.5%) had short segment aganglionosis of the colon.

The most favoured method for the definitive operation in the treatment of Hirschsprung's disease was the Swenson's procedure (52.6%) while only one patient underwent the Duhamel operation.

The commonest preoperative complication seen in these patients was intermittent or complete intestinal obstruction (77.5%).

There were many postoperative complications encountered in the first 30 days, after the definitive operation, the commonest being wound infection accounting for 14.0% and the least common being intra-abdominal or pelvic abscess. One patient died within 30 days after operation in 1994.

Anastomotic stenosis and recurrence of symptoms accounted for 25.9% of the late post-operative complications. They were the reason for re-operations which was done in 21 patients (22.6%). The average postoperative hospital stay was 11 days.

INTRODUCTION

Hirschsprung's disease also called congenital aganglionic megacolon is a congential disorder caused by an abnormal innervation of the bowel resulting in lower intestinal obstruction and chronic constipation. The abnormality in innervation of the bowel extends for varying distances proximally and may involve as little as the lower rectum or as much as the entire colon; in some instances it extends well into the small intestine even to include the entire alimentary canal, exclusive of the stomach [1].

The pathogenesis and pathophysiology of this disease has been contributed to by various studies using electron microscopy, histochemical and immunochemical methods. The presumptive diagnosis of Hirschsprung's disease is usually made through the careful assessment of the presenting history and clinical findings of the patient. Definitive diagnosis of the disease is done by a combination of radiological investigations, rectal manometry and histochemical studies of acetylcholinesterase while confirmation of the disease is by histological examination of biopsies. The definitive treatment of this condition is surgical.

Surgical management of patients with this condition at Kenyatta National Hospital has been changing over the years as in other places. These changes have been attributed to the need of using surgical procedures with fewer complications. At the same time there has been an increase in

the number of surgical specialists in this field. Before the establishment of the paediatric surgical unit in this institution in 1978 general surgeons performed the original Duhamel procedure. Retrospective studies indicate that the rate of complications was high. With establishment of the paediatric surgical unit, more surgical methods were performed with fewer complications and overall satisfactory results.

LITERATURE REVIEW

BACKGROUND INFORMATION

Harold Hirschsprung presented his classic description of the disease entity that bears his name to the Paediatric Congress in Berlin in 1886 [2, 3]. He described two children who both had classical clinical and anatomical characteristics of the disease. The understanding of the pathogenesis of Hirschsprung's disease took several more decades. That the distal colon was the actual site of the abnormality was initially advanced by Tittel in 1901 who identified an absence of ganglion cells in the distal colon of a child with Hirschsprung's disease [4]. In 1946, Ehrenpreis [5] was the first to appreciate that the colon became secondarily dilated because of distal obstruction. In 1948, Whitehouse and Kernohan[6] definitively documented the absence of ganglion cells of the myenteric plexus in patients with Hirschsprung's disease. In 1948 Swenson and Bill published the first results of abdominal 'pull-through' procedure to resect the undilated but aganglionotic distal bowel, leaving the 'megacolon' to revert back to normal after this functional obstruction has been removed. An excellent review of the history can be found in other literature[7].

EPIDEMIOLOGY

Incidence

The incidence of Hirschsprungs disease ranges from 1 in 4400 to 1 in 7000 live births [8,9]. The male to female ratio is generally reported as 4:1 in favour of males [10, 11]. In the long segment disease the ratio approaches 1:1 and may actually become reversed. The age of onset of this disease ranges from soon after birth through childhood.

Risk factors

The risk of Hirschsprung's disease increases in familial cases to approximately 6% with a range of 2% to 18% [13]. Badner et all [14] in their study showed that brothers of patients with short-segment of the disease have a higher risk (4%) than sisters (1%). In long segment Hirschsprung's disease, brothers and sons of affected males have the greatest risk of being affected (24% and 29% respectively). It has also been shown than the prevalence of Hirschsprung;s disease is higher in whites than in blacks.

Associated Congenital defects

The table below summarizes the congenital defects, which have been associated with Hirschsprungs disease.

TABLE: 1 ASSOCIATED CONDITIONS	
Central Nervous system	• Down's syndrome • Occipital encephalocele
Cardiovascular	• Congenital heart disease
Genito urinary	• Hypospadias • Hydronephrosis • Absent kidney • Megacystis • Undescended testis
Gastro intestinal	• Volvulus neonatorum • Pyloric stenosis • Meckel's diverticulum • Anorectal malformation • Mesenteric defect
Hernia	• Inguinal hernia/hydrocele • Umbilical hernia • Oesophageal hiatus hernia
Musculo skeletal	• Talipes equinovarus • Hemivertebrae
Others	• Cataract • Strabismus • Cleft palate • Scalp heaemagioma

PATHOGENESIS

Background knowledge/Embryology and Etiology

In normal embryonic development, neuroenteric cells migrate from the neurocrest to the upper end of the alimentary tract and then proceed in a distal direction [14]. The first cells arrive in the oesophagus by the fifth week and migration to the distal colon is achieved by the twelfth week. The migration occurs first into the myenteric plexus, and these cells subsequently move into the submucus plexus. Neurocrest cells are guided in their migration by various neural glycoproteins. These include fibronectin and hyaluronic acid, which create a pathway for neural migration [15].

Two basic theories regarding embryomic defect in Hirschsprung's disease exist. These are failure of migration, hostile environment and immunologic theories [16,17,18].

Pathology

The gross features of the disease vary with the duration of untreated disease. The intestine may appear fairly normal in the neonatal period but as the child ages, the proximal ganglionic intestine hypertophies and becomes thicker and longer than normal. The taeniae disappear and longitudinal muscle layer seem to completely surround the colon. The transitional zone may be funnel like and vary in length. The distal intestine appears normal

The hallmark of the disease is the absence of ganglion cells in the distal intestine both in the submucosal plexus and the intermuscular plexus. There is marked increase in preganglionic nerve fibres. The aganglionosis process is nearly always continous and uninterrupted. Approximately 80% of aganglionosis typically extend to the rectosigmoid colon [19,20]. The table summarises the incidence of various levels of aganglionosis.

TABLE 2: EXTENT OF AGANGLIONOSIS IN HIRSCHSPRUNG'S DISEASE	
Transitional zone	Percentage
Rectosigmoid	70-75%
Long segment of intestine	15-20%
Total colon	10%
Small intestine	1-10%

Various neuronal and peptide makers that help to identify alteration in ganglionic distribution in Hirschsprung's disease have been identified. Staining of vasoactive intestinal peptide (VIP) – containing nerves, substance P, gastrin releasing peptide, met-encephaline have been shown to be decreased [7,21,22,23]. Neuropeptide Y has been shown to be increased in a ganglionic colon [24]. These changes are yet to be explained.

Pathophysiology

The intestine contains three neuronal plexi; the submucosal, the intermuscular and much smaller mucosal plexus [24]. Each contains a

finely intergrated neuronal network that acts to control all functions of the gut with relatively little control from the body's central nervous system. Normal intestinial motility is primarily controlled by these intrinsic neurones in each ganglion. Extrinisic control of the intestine is through both preganglionic and post ganglionic cholinergic fibres. Cholinergic fibres result in contraction through the neurotransmitter acetylcholine while adrenergic fibers are predominantly inhibitory and use norepinephrine to mediate their function. Bult et al [25] showed that there exists a non cholinergic non adrenergic nervous system in the intestine which is predominately inhibitory and the neurotransmitter responsible for this was nitric oxide (NO).

With the absence of ganglion cells, the extrinsic nervous system develops a markedly increased inervation of the intestine. Adrenergic and cholinergic systems show a two-to-three fold increase in inervation in the aganglionic intestine [26,27]. This led to the conclusion that the spastic contraction of the aganglionic segment was due to:-

(i) Predomination of adrenergic excitatory system over the inhibitory function in the aganglionic segment.

(ii) Marked increase in the predominately excitatory cholinergic inervation in the aganglionic tissue.

(iii) Loss of the intrinisic enteric inhibitory nerves, a nervous system whose major mediator is nitric oxide (NO) [28].

DIAGNOSIS

Diagnosis of Hirschsprung's disease depends first on the clinical picture described above. Barium enema usually confirms this. Anorectal manometry helps in certain cases, and rectal biopsy should clinch the diagnosis.

Clinical presentation

(a) **Newborn:** The usual presentation of Hirschsprung's disease in newborns consist of:-

- A history of delayed passage of meconuim more than 48 hours post delivery
- Newborn constipation and abdominal distension
- Poor weight gain
- Poor feeding
- Emesis
- Rectal examination reveals a normal anal tone and an empty ampulla.

(b) **Infants and children:** Clinical signs include:-

- Chronic constipation with absence of faecal soiling
- Difficulty controlling bowel movements
- Intermittent vomiting
- Large faecal mass on abdominal palpation
- pellet-like stool
- Rectal examination reveals normal anal tone, empty ampulla and explosive evacuation of faecal fluid and gas on withdrawal of finger from the rectum.

(c) A child with Hirschsprungs disease may also present with complications of the disease which include:-

- Intermitent or complete intestinal obstruction with abdominal distension and bilious vomiting
- Toxic megacolon, diarrhoea lethargy, fever, dehydration and/or sepsis indicating the development of enterocolitis
- Failure to thrive
- Protein-loosing enteropathy with hypoproteinemia and oedema or anasarca

- Obstructive uropathy secondary to ureterovesical obstruction

- Melena stool

Imaging studies

(a) Abdominal X-ray: this is taken in either antero-posterior or decubitus abdominal position. Usually it shows the following features:-

- Several distended loops of bowel

- Absence of the normal rectal gas pattern

(a) Barium enema: classical findings include:-

- A spastic distal aganglionic intestinal segment

- Dilated proximal ganglionic segment

- Funnel shaped transitional zone

- Retained barium 24 hours after a barium enema

Barium enema in the first several days or weeks of life may fail to show a transitional zone. Swenson [29] noted an accuracy of 76%, Lister [20], 87%, and Klein [30] 92%.

Anorectal Manometry

This was introduced by Schuster 1965 [31]. This method is based on the fact that in the normal child, the intraluminal pressure of the anal canal falls when the rectum is distended artificially. This phenomenon is thought to be due to the relaxation of the internal anal sphincter muscles and is called rectosphincteric reflex. In Hirischsprung's disease, however, the pressure in the anus rises when the rectum is distended. The accuracy of the method increases greatly with age of the child, as has been demonstrated by Meunier 1978 [32] in his three-year study period when he performed manometric examinaiton on 229 children.

Rectal Biopsy

This is the gold standard for the diagnosis of Hirschsprung's disease. It was first described by Swenson [33] in 1959. Rectal biopsy is performed using either a Dobbin (1965)[34] rectal suction biopsy which is a submucosal biopsy method or a full-thickness rectal biopsy which was described by Kodawaki (1979)[35] in which the identification of ganglion cells is carried out in the submucosal and intermuscular plexus. The latter is the main area of the demonstration of ganglion cells. The technique of suction rectal biopsy is quite safe, although complications have been described. In a review of 134 consecutive biopsies at one institute, three clinical perforations (0.2%) and three haemorrhages requiring transfusion occurred [36]. The accuracy of the test has been well demonstrated. In one of the largest reviews on the use of suction rectal biopsy. Andrassy [37] found only one false negative result in a study of 444 patients. Laparatomy and serial biopsies is the method which has

been carried out for the confirmation of the diagnosis and demonstration of the extent of the aganglionic segment [1].

The following histological features are seen from the rectal biopsies in Hirschsprung's disease:-

- Absence of submucosal and myenteric ganglion cells
- Absence of Meissner's and Aurbach's plexi
- Hypertrophied nerve bundles with high concentration of acetylcholinesterase
- The aganglionic region may involve the transitional segment
- Normal distribution or proliferation of ganglion cells in the region of the colon proximal to the transitional zone.

Differential Diagnosis of Hirschsprung's disease

These include: -

(a) Mechanical obstruction

- Meconium ileus
- Distal ileal or colonic atresia
- Small intestinal stenosis

- Anorectal malformation

(a) Functional obstruction of the intestinal tract

- Prematurity
- Small left colon syndrome
- Meconium plug syndrome
- Sepsis and electrolyte imbalance
- Hypothyroidism
- Functional constipation
- Intestinal neuronal dysplasia

MANAGEMENT

Therapeutic options for Hirschsprung's disease have gradually become refined through trial and error. The first treatment consisted of a diverting colostomy, which would relieve the child's symptoms but symptoms would retum after closure of the colostomy. Attempts at bypass or removal of the redundant portions of the colon were uniformly unsuccessful. One of the more intriguing approaches was a lumbar sympathectomy[38,39]. Theoretically, removal of the sympathetic input to the distal rectum would result in predominance of parasympathetic or relaxation impulse. Several patients noted improvement in symptoms with this approach which was discussed by Ladd and Gross in their 1941 textbook on Paediatric surgery[40].

The first successful surgical management of Hirschsprung's disease was described by Swenson and Bill (1948)[41]. The patients were initially treated with a colostomy. The definitive step of the procedure was an abdomino-perineal rectosigmoidectomy. This procedure included a meticulous extensive pelvic dissection, resection of the aganglionic colon and a pull - through of the normal colon downwards and a two layered colorectal anastomosis near or including the internal sphincter. These authors reported good results. Subsequent modifications of this operation by Swenson involved a more oblique anastomosis and sparing of the internal sphincter. 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. These included retraction or necrosis of the pulled through intestine, leaking anastomosis, intestinal

obstruction, intestinal fistula, pelvic abscess formation and incontinence of stool.

If a child older than 6 months of age is diagnosed having Hirschsprung's disease and is in good health, the Swenson procedure can be performed as a primary procedure eliminating the need for a preliminary colostomy [39].

A simpler but less effective method, that did not resect sufficient lengths of aganglionic bowel was the State procedure[42] which consisted of a low anterior resection of the rectosigmoid colon, this technique has been abandoned. A subsequent refinement was described by Rehbein in 1953, this technique consisted of a low anterior resection of aganglionic intestine, which extended distally well beyond the peritoneal reflection. The results of this procedure have been fairly good and it is still successfully used in many parts of the world [43].

Duhamel [44] and Soave[45] subsequently developed the two techniques that are commonly used today. Recently laparascopically assisted techniques have been developed[46,47].

The Soave or endorectal pull-through technique was introduced by Franco Soave[45] at the Institute G.Gaslini in 1963. This procedure consisted of removing the mucosa and submucosa of the rectum and pulling ganlionic intestine through the aganglionic muscular cuff. The initial procedure was done without a formal anastomosis and relied on scarification over time of the two limbs of intestines to join. The procedure was modified by Boley [48] who performed a primary anastomosis at the dentate line and then further modified the procedure by everting the submucosal – mucosal tube onto the perineum to facilitate

the performance of the anastomosis [49]. The procedure is now commonly performed as a primary pull-through procedure in neonates without the need for an initial levelling colostomy (single/one stage) [50]. The complication rate with the single stage approach is identical to that seen with the standard two-staged approach. The endorectal dissection in the newborn is technically easier to perform than in older children. The endorectal pull through technique avoids injury to the pelvic nerves. Important sensory fibres and the integrity of internal sphincter are preserved. The operation has conventionally been performed on children at 6 to 12 months of age.

The Duhamel operation was first described by Bernard Duhamel in 1956 as a modification of the Swenson procedure. A retrorectal approach was used, and a significant portion of the aganglionic rectum was preserved. The rationale for this procedure includes ease in performance, prevention of anastomotic leak and strictures while eliminating the obstruction and preservation of sensory receptors. The operative principles of this technique include minimal pelvic dissection, a wide but not sutured anastomosis between the ganglionated colon and excluded rectum, a retrorectal approach for the pull-through intestine to the anal canal, division of the internal anal sphincter, and preservation of the anterior wall of the rectum and its nerve supply [44.51]. In his early series Duhamel reported a decrease in complications when compared with the contemporary reports of the Swenson's operation [44,51].

There have been numerous modifications of the Duhamel procedure. Most modifications have centred around the elimination of the common wall and the rectal pouch "spur" and the influence of a stool filled noneliminating blind rectal pouch with an aganglionic rectal wall. Martin

and Alternuer[51] and Martin and Caudil[53] described careful clamp placement to entirely eliminate the rectal pouch with application of mechanic stapling devices to the colorectal anastomosis, the division of the common rectal wall was further facilitated as reported by Ikeda[54], Soper[55] and Ravitch[56].

Total colonic agnaglionosis with or without involvement of the small bowel fortunately is a rare form of Hirschsprung's disease and accounts for 5-8%. Signs and symptoms of total colonic 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 techniques [56,58]. Aganglionosis of the entire colon may be managed surgically by the following approaches.

1. Total colectomy and endorectal ileal pull-through[59].

2. The Martin's procedure where ganglionated small intestine is anastomosed side to side for a variable length with the agangalionic rectum, sigmoid and descendidng colon[60] or

3. Akimura – Stringel right patch enteroplasty [60, 61].

For ultra short aganglionic segment, anorectal myectomy has given good results as reported by Scobie[63]. Anorectal myectomy which deals with spastic internal sphincter and also the propulsive failure of the terminal segment of the rectum has generally gained acceptance as a satisfactory method of treatment in an ultra-short aganglionic segment of Hirschsprung's disease.

The choice of surgical procedure is therefore broad and is entirely at the personal preference of the individual surgeon, namely, the one that gives him the best result.

The common postoperative complications seen after various operations for the treatment of Hirschsprungs disease includes, enterocolitis, anastomotic strictures, adhesive intestinal obstruction, severe perineal excoriation, anastomotic leak, fever above 38°C for more than 3 days, wound infection, ileus (delayed bowel function >5days), bleeding, intra abdominal or pelvic abscess, wound dehiscence among others.

STATEMENT OF RESEARCH PROBLEM

Justification of the Study

Before the early 1970's Hirschsprung;s disease was thought to be a rare disease in the tropics and its medical awareness was correspondingly low. In 1990 a study on Hirschsprung's disease at Kenyatta National Hospital was carried out by Dr. Svetlana M. Barrack. In this study it was shown that the disease was not rare in this country with an average of eleven new cases registered yearly in the institution at that time. The study also outlined the diagnostic and treatment methods which were in place at that time. Ten years down the line, there has been tremendous improvement in the management of this condition. Better diagnostic methods e.g suction rectal biopsy have become a routine in this institution. More surgeons have been trained in Paediatric Surgery. Some old operative methods have been discarded and new ones put in place. It is in this regard that it is expected that better results have been registered in the management of this condition in these last ten year – because of this a study to review the cases of Hirschsprung's disease will highlight the advancement achieved in the management of this disease in this institution.

STUDY OBJECTIVES

Broad Objectives

To review the presentation and management of Hirschsprung's disease at Kenyatta National Hospital in the last ten years.

Specific Objectives

(a) To study the age at presentation and diagnosis of Hirschsprung's disease

(b) To study the clinical presentation of Hirschsprung's disease

(c) To study the diagnostic methods for Hirschsprung's disease

(d) To evaluate the treatment methods for the disease

(e) To study the incidence of complications of the disease and treatment

MATERIALS AND METHODS

Study Design

This was a ten year retrospective study covering the period January 1991 to December 2000 at Kenyatta National Hospital.

Study subjects were identified from the main theatre operation register and their case notes retrieved from Medical Records Department with the assistance of 2 resident clerks.

Relevant data was then extracted from the case notes using a pre-designed proforma questionnaire (a specimen of which is annexed as appendix 1) by the author.

The obtained data was then analysed by computer and is presented in tabular and text form.

Minimisation of errors and biases

An eligibility criteria was adopted. The patients included in the study were:

(a) all those who were attended to at Kenyatta National Hospital with a histological diagnosis of Hirshsprung's disease.

(b) All those who had the definitive surgical procedure performed in Kenyatta National Hospital and had the primary procedure (colostomy) done outside of the institution.

(c) All those who have gone through all the stages of surgery and have been followed up for a period not less than six months from the date of completion of surgical treatment.

The patients excluded from the study were:

a) All those who did not undergo the definitive surgical procedure in KNH and were being followed up in the institution.

b) All those whose colostomies were still open.

c) All those who were lost to follow-up not more than six months after completion of all the stages of surgical treatment

d) All those whose records at KNH were either incomplete or unavailable

Ethical Consideration

The selection was anonymous and retrospective and carried out in strict confidence by the author after approval by the Kenyatta National Hospital Ethics and Research Committee.

Constraints and Weakness of the Study

This being a retrospective study relied heavily on pre-recorded information, which may have been inaccurate, incomplete or omitted. Some of the patients' records were missing from the shelves.

RESULTS

A total of 93 patients underwent surgery for Hirschsprung's disease between January 1991 to December 2000. All were operated upon by surgeons from the paediatric surgical unit of KNH.

Sex Distribution

There were 75 males (80.6%) and 18 females (19.4%) giving a male preponderance of 4.2:1. The sex distribution of the patients is shown in table 1 below.

Table 1: Sex Distribution of Patients Operated on for Hirschsprung's Disease at KNH

SEX	NO. OF PATIENTS	PERCENTAGE %
Male	75	80.6
Female	18	19.4
Total	93	100

The highest number of patients operated in a year was 16 (17.2%) in the year 2000 while the lowest was 4(4.3%) in 1994. There was a steady rise in the number of patients operated each year from 6 in 1991 to 16 in the year 2000. Table 2 below summarises the yearly distribution of patients operated upon.

Table 2 Yearly Distribution of Patients Operated at KNH

Year	No. of Patients	Percentage %
1991	6	6.5
1992	5	5.4
1993	7	7.5
1994	4	4.3
1995	7	7.5
1996	9	9.7
1997	12	12.8
1998	13	14.0
1999	14	15.1
2000	16	17.2
Total	93	100

Age at Diagnosis

The youngest patient who had a histological diagnosis was 14 days old while the oldest was 13 years. Only 5 patients (5.4%) had a histological diagnosis made below the age of 1 month. Most of the patients had histological diagnosis made in the age group 1-5 years (50.4%). Table 3 below summarises the age at diagnosis of the patients operated upon in KNH.

Table 3 Age at Diagnosis in KNH

Age	No. of Patients	Percentage (%)
0 - 1 month	5	5.4
1 - 6 months	14	15.1
6 - 12 months	10	10.8
1 - 5 years	47	50.4
> 5 years	17	18.3
Total	93	100

Age at Operation

The youngest patient was operated at the age of 2 months while the oldest was operated on at the age of 15 years. Majority of patients operated were in the age group of 1-5 years (59.1%). The age group of 6-12 months constituted the least number of patients operated upon. Table 4 below summarises the age at operation of patient with Hirschsprung's disease in KNH.

Table 4 Age at Operation in KNH

Age	No. of Patients	Percentage (%)
1 - 6 month	7	7.5
6 - 12 months	5	5.4
1 - 5 years	55	59.1
5 - 10 years	20	21.5
> 10 years	6	6.5
Total	93	100

Symptoms at Presentation

The symptoms at presentation to hospital are shown in table 5. The predominant presenting features were abdominal distension seen in 91 patients (97.8%), failure to pass meconium within 48 hours in 46 patients (49.5%) and visible peristalsis in 37 patients (39.8%). The least dominant features were enterocolitis and feacal soiling seen in 1 patient each. These were patients who presented in the age group above 5 years.

Table 5 Symptoms at Presentation to Hospital

Symptom	No. of Patient	Percentage (%)
Failure to pass meconium within 48 hours after birth	46	49.5
Abdominal distension	91	97.8
Visible peristalsis	37	39.8
Diarrhoea	8	8.6
Enterocolitis	1	1.1
Failure to deaficate	6	6.5
Constipation	80	86.0
Palpable abdominal mass	9	9.7
Faecal soiling	1	1.1
Failure to thrive	16	17.2

Methods of Diagnosis

Barium enema radiography was diagnostic in 65 patients (69.9%) in this series. Histological diagnosis of Hirschsprung's disease was determined by open biopsy in 70 patients (75.3%) and by suction biopsy in 14 patients (15.1%). However it was noted that suction biopsy was available in the hospital from the year 1997. In this series anorectal manometry and histochemical studies were not carried out in the institution. Table 6 below shows the methods of diagnosis which were used in the diagnosis of the disease in KNH.

Table 6 Method of Diagnosis in KNH

Method	No. of Patients	Percentage (%)
Plain abdominal x-ray	13	14.0
Barium enema Radiography	65	69.9
Laparatomy and serial biopsy	70	75.3
Rectal biopsy	30	32.3
Suction biopsy	14	15.1
Anorectal Manometry	0	0
Histochemical studies	*0*	*0*
Abdominal ultrasound	1	1.1

Level of Aganglionosis

In this series most patients had short segment level of aganglionosis, 87 patients (93.5%) while only 6 patients (6.5%) had long segment of aganglionosis . Of the short segement aganglionosis 54 patients (58.0%) had recto-sigmoid aganglionosis. In patients with long segment aganglionosis, the level of aganglionosis was restricted to the descending and transverse colon. Table 7 summarizes the level of aganglionosis as seen in patients treated at KNH.

Table 7 Level of Aganglionosis in patients treated in KNH

Level of Aganglionosis	No. of Patients	Percentage (%)
Short Segment		
Rectum	19	20.4
Recto sigmoid colon	54	58.0
Sigmoid colon	6	6.5
Unspecified length	8	8.6
Long segment		
Descending colon	4	4.3
Transverse colon	2	2.2
Total	93	100

Definitive Procedure

In this series Swenson's procedure was performed in 49 patients (52.6%), Soave-Boley (staged) procedure in 25 patients (26.9%) and One stage Soave-Boley in 9 patients (9.7%) while only 1 patient underwent the Duhamel procedure. The definitive procedures which were carried out are shown in table 8.

Table 8 The Definitive procedures performed

Procedure	No. of Patients	Percentage (%)
Duhamel	1	1.1
Soave	3	3.2
Soave-Boley (staged)	25	26.9
Soave-Boley (one stage)	9	9.7
Swenson	49	52.6
Bianchi myectomy	6	6.5
Total	93	100

Pre operative complications

Seventy-two patients (77.5%) had intermittent or complete intestinal obstruction at presentation to hospital.

The second most common complication at presentation was failure to thrive which was present in 33 patients (35.5%).

The least common preoperative complication seen in patients was melena stool. Table 9 summarises the rates of preoperative complications.

Table 9 Preoperative complications at presentation to hospital

Complication	No. of Patients	Percentage (%)
Intermitent or complete intestinal obstruction	72	77.5
Enterocolitis	4	4.3
Failure to thrive	33	35.5
Obstructive uropathy	2	2.2
Melena stool	2	2.2

Early post operative complications

In this study there were many postoperative complications seen in the first 30 days after the definitive procedure. The most common was wound infection seen in 13 patients (14.0%).

The least common was pelvic abscess, which was seen in 1 patient (1.1%).

One patient died within 30 days of operation. However the cause of death was not clearly defined since post mortem was not carried out on the body.

Table 10 summarizes the rate of complication seen in the first 30 days after the definitive procedure.

Table 10 *Early postoperative complications (Within 30 days of the definitive procedure)*

Complications	No. of Patients	Percentage (%)
Enterocolitis	3	3.2
Adhesive intestinal obstruction	2	2.2
Severe perianal excoriation	3	3.2
Wound infection	13	14.0
Ileus (Delayed bowel function > 5 days)	2	2.2
Bleeding anastomotic site	3	3.2
Intra-abdominal or pelvic abscess	1	1.1
Mortality (within 30 days of operation)	1	1.1

Late postoperative complications

In this series late complications were defined as those complications seen after 30 days from the date of definitive procedure. The most common late complication was anastomotic stenosis seen in 18 patients (19.4%) followed by intestinal obstruction seen in 15 patients (16.1%) after

colostomy closure. The least common late complication was enterocutaneous fistula seen in 2 patients (2.2%).

Table 11 summarises the late postoperative complications seen in patients operated upon in KNH.

Table 11 Late postoperative complications

Complication	No. of Patients	Percentage (%)
Intestinal obstruction	15	16.1
Postoperative enterocolitis	5	5.4
Incontinence of stool	9	9.7
Fecal soiling	8	8.6
Anastomotic stenosis	18	19.4
Enterocutaneous fistula	2	2.2
Recurrence of symptoms	6	6.5

Rate of Reoperations

There were a total of 21 patients who underwent reoperation. Reoperations were done because of recurrence of symptoms and anastomotic strictures. The year 1991 and the year 1994 recorded the highest rates of reoperation, which was 50%. There was however a general trend in reduction of the rates of reoperation from the year 1991 to the year 2000 as summarized in table 12 below.

Table 12 Yearly rate of Reoperation in KNH

Year	No. of operations	Re-operation	Percentage (%)
1991	6	3	50.0
1992	5	1	20.0
1993	7	2	28.6
1994	4	2	50.0
1995	7	1	14.3
1996	9	1	11.1
1997	12	3	25.0
1998	13	4	30.8
1999	14	2	14.3
2000	16	2	12.5
Total	93	21	22.6

Rate of Reoperations for the Definitive procedure

Out of 49 patients who underwent the Swenson's operation, 11 (21.9%) were re-operated, 3 due to recurrence of symptoms and 6 due to anastomotic strictures.

Eight of the 25 patients (32.0%) who underwent the staged Soave-Boley operation were re-operated, 2 due to recurrence of symptoms and 6 due to anastomotic strictures.

Two of the 6 patients (33.3%) who underwent Bianchi myectomy were re-operated, both of them due to recurrence of symptoms.

The rate of re-operation after each definitive procedure are shown in table 13.

Table 13 Rate of reoperation for the definitive procedure

Procedure	No. of patients operated	Re-operation	Percentage (%)
Duhamel	1	0	0
Soave	3	0	0
Soave-Boley (Staged)	25	8	32.0
Soave-Boley (One stage)	9	0	0
Swenson	49	11	22.4
Bianchi Myectomy	6	2	33.3

Postoperative Hospital stay

The overall average post operative hospital stay in this series was 11 days.

Table 14 summarizes the yearly average postoperative hospital stay.

Table 15 shows the average postoperative hospital stay seen after the various definitive procedures carried out. The shortest postoperative hospital stay was 1 day, after Bianchi myectomy while the longest stay was 32 days. The patient who stayed for 32 days had pelvic abscess. The mean postoperative hospital stay was shortest after Bianchi myectomy at 3.4 days, and longest after the Soave procedure at 23.7 days.

Table 14 Yearly average postoperative hospital stay.

Year	No. of Operations	Mean Post-op Hospital Stay	Standard Deviation (STD Dev.).
1991	6	11.8	0.80
1992	5	9.8	4.1
1993	7	11.4	2.5
1994	4	14.3	3.6
1995	7	12.3	6.5
1996	9	14.4	8.9
1997	12	10.7	3.9
1998	13	11.6	3.9
1999	14	9.0	2.3
2000	16	9.0	2.7
For entire population	93	11.0	4.4

Table 15 *Average postoperative hospital stay for the definitive operations performed*

Operation	No. of patients	Mean	STD Dev.
Duhamel	1	8.0	
Soave	3	23.7	9.5
Soave-Boley (staged)	25	11.0	3.1
Soave-Boley (one stage)	9	9.8	2.7
Swenson	49	11.2	3.1
Bianchi Myectomy	6	3.4	3.2
For entire population	93	11.0	4.4

DISCUSSION

Hirschsprung's disease is a relatively common disease. World wide the male to female ratio in patients with classic Hirschsprung's disease is generally reported as 4:1 [58]. In this series it is in conformity with what is generally seen the world over.

The median age at which children are diagnosed with Hirschsprung's disease has progressively decreased over the past several decades from 2 to 3 years of age during the first decades of this century to a mean of between 3 to 6 months of age during the 1950s to 1980s [20].

In this study most children (50.4%) were diagnosed with the disease between the ages of 1 to 5 years. However, in Kenya, Kenyatta National Hospital is the only centre were operations for Hirschsprung's disease are carried out. The delay in diagnosis is partly contributed to by delayed referrals from the peripheral health institutions where the children first present. It was not possible to determine at what age the children first presented to a health institution because of lack of and/or un -availability of clear and detailed referral notes.

In this series it was also noted that most children with the disease (59.1%) were operated upon between the age of 1 to 5 years. This therefore means that though there was overall delay at the age of diagnosis, the children were operated upon soon after the diagnosis was made. There was general increase in the number of children operated upon from 6 in

1991 to 16 in the year 2000. In the first 5 years of this study (1991 - 1995) an average of 6 children were operated upon yearly while in the last 5 years (1996 - 2000) an average of 13 children were operated upon yearly. This rise in the number of children operated yearly was attributed to by the high number of paediatric surgeons who joined the paediatric surgical unit in the last 5 years of the study.

The usual presentation of Hirschsprung's disease in newborns consist of a history of delayed passage of meconium within 48 hours of birth. This is seen in about 95% of full term infants with the disease [50]. In this series this presentation was seen in only 49.5% of the patients. Other presenting signs which are commonly seen including constipation, abdominal distension and failure to thrive were all common features in this study.

Barium enema radiography has been shown to have good rates of diagnostic accuracy of upto 92% [34]. In this study this diagnostic method was used in 69.9% of patients. It's a procedure with minimal morbidity.

Rectal biopsy was thought to be the gold standard for diagnosis of Hirschsprung's disease [20].

Refinement of this technique led to the development of suction rectal biopsy by Dobbins and Bill [7,41]. Although the technique of suction rectal biopsy is quite safe, it was noted that only 14 patients (15.1%) underwent this procedure despite the fact that the procedure was introduced in this hospital in 1997. The pathological evaluation of a suction rectal biopsy for the diagnosis of Hirschsprings disease is facilitated by

acetylcholinesterase stain yet in this series this staining technique was not used at all.

Anorectal manometry was not used in the diagnosis of Hirschsprung's disease. This method of diagnosis has the advantage of being able to be done at the bedside or as an outpatient procedure as it is associated with virtually no complications [30].

Laparatomy and serial biopsy has the advantage of decompressing the gut and also giving a biopsy specimen for histological diagnosis of the disease. In this study this method was widely used in 70patients (75.3%).

In this study, 87 patients (93.5%) had short segment aganglionosis while only 6 patients (6.5%) had short segment aganglionosis. Rectosigmoid aganglionosis constituted 58.0% of the total number of patients seen. The pattern of level of aganglionosis observed in this study is in conformity with what other studies have shown [57].

Swenson's procedure was the most favoured procedure in this series. Fouty nine patients (52.6%) underwent the Swenson's procedure, 25 patients (26.9%), the staged Soave-Boley procedure and 9 patients (9.7%) the one stage Soave-Boley procedure. The least favoured procedure was the Duhamel procedure, 1 patient and Soave procedure 3 patients. Six patients underwent Bianchi myectomy procedure. These were patients with what was thought to be ultra short segment aganglionosis.

Intermitent or complete intestinal obstruction were the most common preoperative complications seen in 72 patients (77.5%). Failure to thrive was seen in 33 patients (35.5%). This was expected since most patients were diagnosed at late age of between 1 to 5 years.

Although the early postoperative complications were many their rate of occurrence were low with the highest being wound infection which was seen in 13 patients (14.0%).

Twenty-one patients (22.6%) were reoperated upon. 12 patients out of these had anastomotic stricutre which did not improve on dilatation. 6 patients had anastomotic strictures which improved on dilatation. 5 patients who were reoperated had recurrence of symptoms and on further evaluation were found to have residual aganglionotic segment after closure of colostomy. In this series it was also noted that none of the patients who underwent one stage Soave-Boley procedure was reoperated upon. Overall, staged Soave-Boley operation had the highest rate of reoperation at 32.0% of the patients who underwent this procedure.

Although the average postoperative hospital stay was 11 days, there was a fall in the mean post operated hospital stay from periods of over 10 days in the earlier year to 9 days in 1999 and in the year 2000. However it was noted that many patients who were well enough to go home were kept in the ward awaiting a digital rectal examination on the 10^{th} postoperative day.

CONCLUSIONS AND RECOMMENDATIONS

Hirschsprung's disease is common in this country. Currently Kenyatta National Hospital is the only public hospital in this country that has the capability to treat patients with these conditions.

Although children with this condition present to a health facility in infancy the definitive, histological diagnosis is made late and therefore it is recommended that many doctors, nurses and other support staff be trained in this field and be dispatched to other peripheral hospitals at least in the level of provincial hospital to manage this condition and other paediatric surgical conditions.

Suction rectal biopsy being a safe, and accurate procedure should widely be used in the diagnosis of the disease since this procedure is now available in this institution. Similarly anorectal manometry should be made available to make easier the diagnosis of ultra short segment aganglionosis, which may be a major cause of recurrence of symptoms in unsuspected cases.

Where the diagnosis has been made early and the child is suitable for the procedure, one stage Soave-Boley endorectal procedure should be carried out since this methods allows the child to leave hospital early and saves the child the trauma of a second and probably a third stage operation.

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APPENDIX 1

QUESTIONNAIRE/DATA COLLECTION FORM

A. DEMOGRAPHIC

Hospital number ……

1. Study number ☐☐
2. Age (Years) ☐☐
3. Sex (male = 1 Female = 2) ☐

B. AGE AT DIAGNOSIS

(Code 0=No 1= YES)

1. 0-1 month ☐
2. 1-6 months ☐
3. 6-12 months ☐
4. 1-5 years ☐
5. >5 years ☐

C. SYMPTOMS AT PRESENTATION

(Code 0=No 1=Yes)

1. Failure to pass meconium ☐
2. Abdominal distension ☐
3. Visible peristalsis of bowel ☐
4. Diarrhoea ☐
5. Vomiting ☐
6. Enterocolitis ☐
7. Failure to defaecate ☐
8. Constipation ☐
9. Failure to thrive ☐
10. Palpable abdominal masses ☐

D. METHOD OF DIAGNOSIS

(Code 0= No 1= YES)

1. Plain abdominal X-ray ☐
2. Barium enema Radiography ☐
3. Laparatomy and open biopsy ☐
4. Rectal biopsy ☐
5. Suction biopsy ☐
6. Anorectal Manometry ☐
7. Histology ☐
8. Histochemical studies ☐
9. Others (specify) ☐

E. LEVEL OF AGANGLIONOSIS

(Code 0=No 1=YES)

Short Segment

1. Rectum ☐
2. Recto sigmoid colon ☐
3. Sigmoid colon ☐
4. Unspecified length ☐

Long segment

1. Descending colon ☐
2. Transverse colon ☐
3. Caecum ☐
4. Total colon and ileum ☐
5. Total colon and jejunum ☐
6. Unspecified length ☐

F. YEAR OF OPERATION

Code 0 = No 1 = YES

1. 1991 ☐
2. 1992 ☐
3. 1993 ☐
4. 1994 ☐
5. 1995 ☐
6. 1996 ☐
7. 1997 ☐

8. 1998 ☐
9. 1999 ☐
10. 2000 ☐

G. THE DEFINITIVE PROCEDURE

(Code 0=No 1=YES)

1. Duhamel ☐
2. Extended Duhamel ☐
3. Soave ☐
4. Soave – Boley (Staged) ☐
5. Soave-Boley (one stage) ☐
6. Rehbein-State ☐
7. Swenson ☐
8. Bianchi myectomy ☐
9. Extended myectomy-myotomy ☐
10. Unknown ☐

H. PREOPERATIVE COMPLICATIONS

(Code 0=NO 1=YES)

1. Intermitent or complete intestinal obstruction ☐
2. Enterocolitis ☐
3. Failure to thrive ☐
4. Protein losing enteropathy ☐
5. Obstructive uropathy secodary to ureterovesical

obstruction ☐

6. Melena stool ☐

I. EARLY POSTOPERATIVE COMPLICATIONS

(Code 0=NO 1=YES)

1. Enterocolitis ☐
2. Anastomotic stricture ☐
3. Adhesive instinal obstruction ☐
4. Severe perianal excoriation ☐
5. Anastomotic leak ☐
6. Fever above 38^0C for more than 3 days ☐
7. Wound infection ☐
8. Ileus (Delayed bowel function >5 days) ☐
9. Bleeding ☐
10. Intra-abdominal or pelvic abscess ☐
11. Wound dehiscence ☐
12. Other complications (specify) ☐

J. DELAYED/LATE POSTOPERATIVE COMPLICATIONS

(Code 0=NO 1=YES)

1. Intestinal obstruction ☐
2. Post operative enterocolitis ☐
3. Anastomotic disruption ☐
4. Faecal incontinence ☐
5. Entero cutaneous fistula ☐

6. Anastomotic stenosis ☐
7. Faecal soiling ☐
8. Mortality ☐

K. AGE AT OPERATION

Code: *0 = No 1 = YES*

1. 0 - 1 month ☐
2. 1 - 6 months ☐
3. 6 - 12 months ☐
4. 1 - 5 months ☐
5. 5 - 10 months ☐
6. > 10 years ☐

L. POST - OP HOSPITAL STAY

Give days ☐☐

M. MORTALITY (Death within 30 days of operation)

Code 0 = No 1 = YES ☐

N: RE-OPERATION

Code 0 = No 1=YES ☐

APPENDIX 2

APPROVAL BY KENYATTA NATIONAL HOSPITAL ETHICAL AND RESEARCH COMMITTEE.

Attached below is a letter of approval of the research proposal "A REVIEW OF THE MANAGEMENT OF HIRSCHSPRUNG'S DISEASE AT KNH (JANUARY 1991 - DECEMBER 2000)" from the Kenyatta National Hospital Ethical and Research Committee.

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Ref. KNH-ERC/01/1229 Date 8th January 2002

Dr. Otieno G. W. Onyango
Department of Surgery
Faculty of Medicine
University of Nairobi

Dear Dr. Otieno,

RE: **RESEARCH PROPOSAL** "A REVIEW OF THE MANAGEMENT OF HIRSCHSPRUNG'S DISEASE AT KNH (JANUARY 1991 - DECEMBER 2000) (P109/10/2001)

This is to inform you that the Kenyatta National Hospital Ethical and Research Committee has reviewed and approved 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

c.c. Prof. K.M. Bhatt,
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