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Management and outcome
Of Hirschsprung’s Disease,
in Soba University Hospital

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وَقَالَ اَللَّهُ ﷺ : ﴿ۖقَلِیلاً إِلَّا الْعَلْمُ مِنَ الْأَوْتُمَّ وَمَا ۗ اَللَّهُ ﷺ ٱلْقَدِیرُ﴾
Dedication

To the soul of my father
To my kind mother
To my brothers, sisters, family
Colleagues
&
friends
without their support neither this nor any professional success would be worthwhile.
Acknowledgements

I acknowledge my gratitude to Mr. Omer Elamin Mohamed Khair, consultant paediatric surgeon for supervising this work with all interest.

I should also thank the Directory of Soba University Hospital for putting the study materials at my disposal. It is also my pleasure to extend my thanks to all the staff members and nursing staff at Paediatric Surgery Unit in S.U.H.

I am also indebted to all my colleagues, the surgical registrars for their moral support. I am indebted to all.
**ABBREVIATIONS**

<table>
<thead>
<tr>
<th>Abbreviation</th>
<th>Meaning</th>
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<tr>
<td>F.H</td>
<td>Family History</td>
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<tr>
<td>G.I.T</td>
<td>Gastrointestinal Tract.</td>
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<td>HD</td>
<td>Hirschsprung’s Disease</td>
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<tr>
<td>MS</td>
<td>Multiple Stage</td>
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<td>SS</td>
<td>Single Stage</td>
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<tr>
<td>S.U.H</td>
<td>Soba University Hospital</td>
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<tr>
<td>U.D.T</td>
<td>Undescended Testicles</td>
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<tr>
<td>V.S.D</td>
<td>Ventricular Septal Defect</td>
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ABSTRACT

Introduction: Hirschsprung's disease (HD) is a congenital disease resulting from failure of caudal migration of ganglion cells in Auerbach’s and Meissner’s plexuses, resulting in intestinal obstruction.

Objectives: To evaluate Sudanese patients with HD, their presentation, diagnosis and treatment.

Methods: A descriptive retrospective and prospective study was performed in 40 patients with HD, at Soba University Hospital, from January 1999 to March 2003.

Data tabulated included personal data, presentation, family and drug history, abdominal examination, investigations, treatment and outcome.

Results: The main symptoms are failure to pass meconium in first 24 hours of life in 36 patients (90%), constipation in 36 patients (90%) and abdominal distension in 38 patients (95%).

Swenson’s procedure was carried in all patients, preceded by a loop colostomy, which was transverse in 7 patients (17.5%), and sigmoid in 33 patients (82.5%).

In 13 patients (32.5%) postoperative complications occurred. Enterocolitis occurred in 5 patients (12.5%) post-operatively, anastomotic stricture in 2 patients (5%), wound infection in 7 patients (17.5%) and stool incontinence in 2 patients (5%).

Death occurred in 2 patients (5%), both developed postoperative enterocolitis, one of them had Down's syndrome.

Conclusion: The treatment of HD is surgery and can be done early.
الخلايا هجرة في القصور وفقاً لوالديه. داء هرشبرنغ، داء وعائمة للأعراض المعروفة في العلاج ونتائج التحليل.


النتيجة: هى الرئيسية للاعراض:

- في العقدية: 24 من 50 مريضاً (90%) في الإمساك، 36 من 50 مريضاً (100%)
- في الرئة: 95 من 100 مريضاً (95%) في تشريح.
- في الحشرة: في الالوان 7 من 50 مريضاً (17.5%) في حادثة جراحية وفغرة 33 من 50 مريضاً (66%)

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INTRODUCTION

Hirschsprung’s disease (HD) “congenital megacolon” is a congenital disease resulting from failure of distal migration of ganglion cells in Auerbach’s and Meissner's plexuses.

The length of bowel affected depends on the level of arrest, the involved segment became collapsed and the proximal part dilated in an attempt to propel faecal material leading to functional intestinal obstruction. Therefore, it is a serious disease may be lethal if not discovered early and treated properly, on the other hand, may present with features typical of fulminating enterocolitis in form of copious offensive diarrhoea.

The management of HD is surgical in a paediatric surgery Department. In the Sudan, the Department of Paediatric in Soba University Hospital (SUH) is one of the main hospitals concerned with this problem. This makes SUH a suitable place to undertake the study on HD. According to my personal communications and literature search no previous local study of the subject was done. This was a good reason for undertaking this study hoping to find out where do you stand.

LITERATURE REVIEW
Anatomy:

Sigmoid colon:

About 10 – 15 inches (25 – 35 cm) long and begins as a continuation of the descending colon in front of the pelvic brim. Below, it becomes continuous with the rectum in front of the third sacral vertebra. Is attached to the posterior pelvic wall by the fan- shaped sigmoid mesocolon. The root of the mesocolon resembles an inverted V. One limb of the V is attached a long the medial side of the left external iliac artery, and the other limb runs downward infront of the sacrum, as far as the third sacral vertebra. At the apex of the V is a small peritoneal recess called the recess of sigmoid mesocolon. Lying beneath the floor of the recess is the left ureter.

Relations:

Anteriorly: In the male, urinary bladder, in the female, the posterior surface of the uterus and upper part of the vagina.

Posteriorly: The rectum and the sacrum, lower coils of the terminal part of the ileum.

Blood supply: Arteries from the sigmoid branchs of the inferior mesentric artery. Veins correspond to the arteries and are tributaries of the portal venous system.

Lymph drainage: Drain into nodes along the course of the sigmoid arteries, from these nodes the lymph travels to the inferior mesentric node.

Rectum:

About 5 inches (13 cm) long, begins in front of the third sacral
vertebra. Passes downward, following the curve of sacrum and coccyx by piercing the pelvic diaphragm and becoming continuous with the anal canal. The lower part of the rectum that lies immediately above the pelvic diaphragm is dilated to form the rectal ampulla.

The puborectalis portion of the levator ani muscles forms a sling at the junction of the rectum with the anal canal and is responsible for pulling this part of the bowel forward, producing the anorectal angle.

The peritoneum covers the anterior and lateral surfaces of the upper third, and only the anterior in the middle third, leaving the lower third devoid of peritoneum.\(^{(1)}\)

The muscular coat of the rectum is arranged in the usual outer longitudinal and inner circular layers of smooth muscles. The three *Teniae coli* of the sigmoid colon, however, comes together, so the longitudinal fibers form a broad band on the anterior and posterior surfaces of the rectum.

The mucous membrane of the rectum, together with the circular muscle layers, forms three permanent folds called the transverse folds of the rectum. These folds are semicircular, two (the upper and the lower) are placed on the left rectal wall and the middle one is on the right rectal wall. \(^{(1)}\)

**Relations:**

- **Posteriorly:**
The rectum is in contact with the sacrum and coccyx, the piriformis, coccygeus, and levators ani muscle, the sacral plexus and sympathetic trunk.

- **Anteriorly:**

  In the male the upper two thirds of the rectum which is covered by peritoneum is related to the sigmoid colon and coils of ileum that occupy the rectovesical pouch. The lower third of the rectum, which is devoid of peritoneum, is related to the posterior surface of the bladder, to the termination of vas deferens and the seminal vesicles on each side, and the prostate. These structures are embedded in visceral pelvic fascia.

  In the female upper two third, sigmoid colon and coils of ileum that occupy the recto uterine pouch (pouch of Douglas) the lower third is related to the posterior surface of the vagina.\(^{(1)}\)

**Blood supply:**

*Arteries:* the arterial supply to the rectum is from the superior, middle, and inferior rectal arteries.

  Superior rectal artery is direct continuation of the inferior mesenteric artery (supply mucous membrane) give left and right branches. The middle rectal artery is a branch of internal iliac artery, it supply the muscular coat. The inferior rectal artery is a branch of internal pudendal artery, it anastomosis with middle rectal artery at level of anorectal junction.

*Veins:*

  Correspond to the arteries. Superior rectal vein (portal circulation) drains into inferior mesenteric vein. The middle and inferior rectal veins drain
into internal iliac and internal pudendal vein respectively, the union between the rectal veins forms porto-systemic anastomosis.

**Lymph drainage:**

The lymph vessels of the rectum drain into a group of nodes embedded in the perirectal connective tissue, just outside the muscular coat, these called pararectal nodes. Lymph vessels draining the upper middle members of this group to the inferior mesenteric nodes. Lymph vessels from the lower part of the rectum follow the middle rectal artery to the internal iliac nodes. The nerve supply (sympathetic and parasympathetic) of the rectum is only sensitive to stretch. (1)

**Anal canal:**

The anal canal is 3 cm long in adult, it is a tube of muscle like the rest of the gut, but the fibres are all circular, consisting of the internal (smooth) and the external (striated) sphincters. It contains no ganglia.

The junction of rectum and anal canal is at the pelvic floor (at level of puborectalis muscle) clasps the gut and angles it forwards. From this right-angled junction with the rectum the anal canal passes downwards and somewhat backwards to the skin of the perineum. (2)

**The lining of the anal canal:**
The canal is lined with mucous membrane in its upper two-thirds, and skin in its lower one-third. The junction of the two is abrupt; it is at Hilton’s white line. This line is the site of attachment of the fascia derived from the longitudinal muscle coat of the rectum, and it is a watershed dividing upper and lower zones of arterial supply and venous and lymphatic return. It also separates two zones of different nerve supply. The part above Hilton’s line (mucous membrane) is derived from the endoderm of the cloaca, while the part below the line (skin) is derived from the ectoderm of the anal pit or proctodaeum.

The cloacal part, is lined with typical large gut mucous membrane containing mucous crypts and covered with columnar epithelium, the lower part lined with squamous epithelium. The arterial supply of the cloacal part is derived from the superior rectal artery, the lower third (skin) below Hilton’s line, is supplied by the inferior rectal artery, which also supplies the sphincters. This arteries do not anastomose with each other.

The veins of the upper part (mucous membrane) drain upwards into the submucous plexus of the ampulla of the rectum, the veins of the skin part, drain, downward, into the inferior rectal vein or into tributaries of the saphenous vein. They do not communicate with the veins of the cloacal part. Hilton’s line is a venous watershed. The lymphatics of the cloacal part passes upwards from the lymphatic follicles in the mucous membrane to join those of the rectum, those of the skin part drain with the rest of
the perineum into the medial group of superficial inguinal nods.

The nerves of the cloacal part are autonomic, from the inferior hypogastric plexuses, relatively insensitive to touch, the sensory innervation comes from $S_2$ and $S_3$ (parasympathetic). The nerves of the skin part are somatic, from the inferior rectal nerve and this skin is highly sensitive. (2)

**Anal sphincters:**

The internal sphincter is of smooth muscle and lies around the upper two-thirds of the canal, the external sphincter is of striated muscle, it clasps the lower part of the internal sphincter and surrounds the lower two thirds of the anal canal.

The internal sphincter is innervated by the inferior hypogastric plexuses, sympathetic stimulation contracts the muscle, parasympathetic stimulation relaxes it. It is under a voluntary control similar to that of the smooth muscle of the bladder, the sphincter is relatively weak, and is not competent when acting alone. External sphincter consist of three parts, It is essential for cleaning of anus after defecation (corrugator cutis anii). (2)

**Large bowel anatomy in the neonate:**

*Large intestine at birth is about 66 cm long*

*and averages 1cm in width, the caecum is*
relatively smaller than in the adult, it tapers into the vermiform appendix. The ascending colon is shorter in the neonate, due to the shorter lumbar region, the transverse colon is relatively long, the descending colon is short, but twice the length of the ascending colon.

The sigmoid colon may be as long as transverse colon, it often touches the inferior part of the anterior body wall on the left and in approximately of neonates part of the sigmoid colon lies in the right iliac fossa. Generally in the colon the muscularies, including the taeniae coli, is poorly developed. Appendices epiploicae and haustra are not present, giving smooth external appearance to the colon.
Haustra appear within the first 6 months. The rectum is relatively long, its junction with the anal canal forms at nearly a right angle.\(^{(3)}\)

**Innervation of the gut:**

The nerve supply of the gastrointestinal tract comprises extrinsic and intrinsic components. The extrinsic components is provided by the autonomic nervous system (parasympathetic and sympathetic) the oesophagus, stomach, small intestine and the proximal colon derive their parasympathetic supply from the vagus nerve. The colon is also innervated by the sacral spinal nerves. The sympathetic supply to the gut is through the lower thoracic and lumbar spinal nerves. The intrinsic innervation is through the submucosal (Auerbach) and myenteric (Meissner) plexus, which contain intercommunicating ganglia.\(^{(4)}\)

**Neural control of gastrointestinal function:**

The gastrointestinal tract has a nervous system all its own called the enteric nervous system. It lies entirely in the wall of the gut, beginning in the esophagus and extending all the way to the anus. The number of neurons in this enteric system is about 10,000,000 almost exactly equal to the number in the entire spinal cord. It controls gastrointestinal movement, and secretion.\(^{(4)}\)

**The enteric system is composed mainly of two plexuses:**

An outer plexus lying between the longitudinal and circular muscular
layer called the myenteric plexus or Meissner’s plexus. An inner called the submucosal or Auerbach’s plexus, that lie in the submucosa.

The sympathetic (mainly postganglionic) and parasympathetic (mainly preganglionic) and sensory fibers connect to the entire nervous system (Myenteric, submucosal plexus).

The myenteric plexus control mainly the gastrointestinal movement, and the submucosal plexus controls mainly local gastrointestinal epithelial secretion and local blood flow.\(^{(4,5)}\)

Different types of neurotransmitters secreted by the Enteric Neuron. Research workers have recently identified a dozen or more different neurotransmitters substances that are released by nerve ending, of different types of enteric neurons.

1. Acetyl choline (excites G.I.T activity).
2. Norepinephrine (inhibit G.I.T activity)
3. Others:
   - Adenosine triphosphat ATP.
   - Serotonin.
   - Dopamine.
   - Cholecystokinin.
   - Substance P.
   - Vasoactive intestinal polypeptide.
• Somatostatin.
• Leu- enkephalin.
• Met- enkephalin.
• Bombesin (their function still to be determined).\(^{(5)}\)

**Hirschsprung’s disease (congenital megacoloon):**

It is due to a congenital absence of ganglion cells in the parasympathetic Auerbach and Meissner complexes of the rectum. The aganglionic rectal segment remains contracted due to unopposed action of the sympathetic nervous system, the bowel above becoming grossly distended and hypertrophied.\(^{(6,7)}\)

**Incidence and genetics:**

The incidence is approximately 1 in each 5000 live births with a 4 : 1 male predominance (short segments) and 1:1 (long segments).\(^{(6,9)}\) HD is a familial disease. Seventeen of 326 index male cases and 13 of 88 index female cases had an affected sibling, with an overall incidence of 3.6% among siblings of all index cases. Consanguinity of parents is exceptional, and only three such instances have been reported in a study of 326 patients. The disease is discordant in dizygotic twins and appears to be concordant in monozygotes.\(^{(10)}\)
Association of congenital aganglionosis of colon with Down’s syndrome (mongolism) is ten times more frequent than would be expected by chance. Approximately 2% of the patients with congenital megacolon have Down’s syndrome. Other anomalies reported to be associated with H.D include megacystis and megaurter, hydrocephalus, V.S.D., cystic deformities of the kidney, U.D.T, diverticulum of the urinary bladder, imperforate anus, Meckle’s diverticulum, hypoplastic uterus, polyposis of the colon, ependymoma of the fourth ventricle, and Laurence- Moon-Biedl- Bardet syndrome.\(^{10}\)

**Pathogenesis:**

Aganglionosis is due to arrest of migration of neural crest cells to the 3 plexi of intestine, under normal circumstances Neural crest cells reach the small intestine in the seventh week of gestation and the rectum in the 12\(^{th}\) week. Thus the aganglionic segment always extend, from the internal anal sphincter for a variable distance proximally. The earlier is the migration arrest, the longer is the distal aganglionic intestinal portion. This migration occurs from cephalad to caudad, and from the midline laterally, which accounts for the greatest distribution of the aganglionic segment in the rectum (30%) and rectosigmoid (44%). Other portions of the intestinal tract are involved less frequently.
Descending colon 11%, splenic flexure 4%, transverse colon 2%, ascending colon 1% and small intestine 8%. Involvement of the entire colon is infrequent, and reported cases of aganglionosis extending proximally throughout the entire small intestine are extremely rare. Thus the hallmark of diagnosis is the absence of ganglion cells in all nerve plexuses. The disease always starts in the rectum and goes proximally with no skip areas. (9,10,11,12)

Morphologically, ganglion cells are absent from the narrowed segment and for a variable distance usually (1 to 5 cm) into the dilated segment. The pattern of nerve fibres is often abnormal also specific stains for catecholamines and cholinesterase have produced variable results, and proposals for the basic pattern of innervation have varied similarly.(10)

Giuseppe Martucciello, et al (2000)(11) in their study about pathogenesis of HD reported, genetic mapping and mutation screening of candidate genes, together with the study of several natural and knockout animal models, clearly have shown the involvement of several different genes in the pathogenesis of HD. Among these genes, the RET proto-oncogene account for the highest proportion of both familial and sporadic cases.

Different animal and human genetic studies have identified 6 Hirschsprung genes: RET proto-oncogene (RET), endothelin 3(EDN3), endothelin B receptor gene (EDNRB), glial-cell-livederived neurotrophic factor (GDNF), endothelin converting enzyme (ECE1), gene encoding the Sry-related transcription factor (so $\times$ 10). Micro environmental factors also can play a role in the pathogenesis of aganglionosis.(11)

Pathophysiology:

The most characteristic abnormality of function in aganglionosis is a failure of relaxation of the internal anal sphincter following rectal distension. In health “a rectal inhibitory reflex” is demonstrable; thus, transient distension of a balloon in the rectum produces a decrease in intraluminal pressure at the level of the internal sphincter, often accompanied by a reflex contraction of the external sphincter.(10)

Although up to 20% of normal children may have a false absence of the reflex, especially if they are premature or of low birth weight, a positive response is a strong evidence against HD,
another pathophysiologic aspect of colorectal motility in aganglionosis is a failure of the contracted segment to relax after parasympathomimetic agents; by contrast, the innervated or dilated segment usually relaxed in aganglionosis and in most healthy control subject, a third abnormality described recently is a (stiff) rectal wall in HD. This increase resistance to stretch was noted even in the dilated, uncontracted bowel, moreover, the greater the degree of “stiffness” the more severe is the clinical picture. Reasons, for these changes are unknown.\textsuperscript{(10)}

Acetyl cholinesterase is present in excessive amounts in the mucousa of the aganglionic segment. The significance of this finding is uncertain, but some have proposed this histochemical approach, which is applicable even to superficial biopsy specimen, as a diagnostic test.\textsuperscript{(10)}

**Clinical picture:**

In most instances, regardless of the eventual severity of the clinical picture, the initial passage of meconium is delayed 48 hours or more after birth in (95\%).\textsuperscript{(9,10)}

The typical presentation of HD consists of failure to pass meconium spontaneously within the first 24 hours of life, followed by constipation, abdominal distension, bilious vomiting, and failure to thrive if not detected in the neonatal period.\textsuperscript{(12)}
The subsequent clinical course varies, some infants continue with complete obstruction do not pass any meconium, and require surgery in the first few days of life. In others, there is recurrent or incomplete obstruction, usually relieved by repeated enemas after the second or third day of life. Other manifestation is development of fulminating enterocolitis with bloody offensive diarrhoea.(10)

**Abdominal examination:**

Digital examination usually reveals no stool in the rectum, but withdrawal of the finger or of a rectal tube may lead to a gush of faeces and flatus with eventual decompression.(10)

One of every four patients with meconium plug, ultimately proves to have congenital megacolon. Vomiting ensues if there is no relief after 48 to 72 hours.(10)

**Abdominal X-ray:**

Although plain abdominal films demonstrate dilated gas and fluid-filled loops, delineation of small and large bowels is difficult in the neonate and the films is often not diagnostic.(10,12)

Barium enema also is not likely to be diagnostic, because sufficient time may not have elapsed in a neonate for the characteristic transitional zone, from constricted to dilated segments, to develop.(10) Barium enema may reveal the sharp
difference in contrast between the dilated normal colon and the aganglionic bowel.\(^{(12)}\)

**Hirschsprung associated Enterocolitis:**

Teitelbaum, *et al.*, (1988)\(^{(13)}\) in their study from 1975 to 1985, 80 infants and children were treated at a major Paediatric Hospital for HD, 19(24\%) of whom developed enterocolitis, they reported a significant risk factors for development of Hirschsprung associated enterocolitis were delay in diagnosis beyond one week of age and the presence of trisome 21.

Elhalaby, *et al.*, (1995)\(^{(14)}\) in their study about enterocolitis associated with HD: a clinical radiological characterization based on 168 patients treated from July 1974 through October 1992. Enterocolitis occurred in 57 patients (33.9\%), either preoperatively 13(7.7\%) or postoperatively 36(21.4\%). In 8 patients (4.8\%) it occurred pre and postoperatively, they report, Hirschsprung enterocolitis present as abdominal distension and explosive diarrhoea associated with the intestinal cutoff sign. The occurrence of explosive diarrhoea in any patient with HD is suggestive of Hirschsprung enterocolitis, even in the absence of systemic symptoms, and should be treated to avoid the morbidity and potential mortality of Hirschsprung enterocolitis.
Acute enterocolitis is the most serious complications of HD, ulceration and necrosis begins in the dilated segment and may extend into the small intestine. Perforation, pericolonic abscess, and septicaemia may occur; and in a more chronic form, anorexia and exudative enteropathy develop. The bowel must be decompressed by a loop colostomy and placed at rest, while fluid and blood are replaced, broad-spectrum antibiotic coverage also is given. Under these circumstance most surgeons advise deferring definitive surgery for aganglionosis for at least six months, to allow complete healing of the colitis.\(^{(10)}\)

**Clinical grading for Hirschsprung associated enterocolitis:**

**Grade I:** mild explosive diarrhoea, mild or moderate abdominal distension, no systemic manifestation.

**Grade II:** moderate explosive diarrhoea, moderate to severe abdominal distension, mild systemic symptoms.

**Grade III:** Severe explosive diarrhoea, marked abdominal distension, shock or impending shock.\(^{(20)}\)

**Pathological grading of Hirschsprung associated enterocolitis:**

**Grade 0:** normal mucous.

**Grade I:** Crypt dilatation, mucin retention.

**Grade II:** Cryptitis or \(\leq 2\) crypt abscesses per high power field.

**Grade III:** multiple crypt abscesses per high power field.
**Grade IV:** Fibrinopurulent debris and mucosal ulceration.

**Grade V:** Transluminal necrosis or perforation.\(^{(20)}\)

The initial presentation in those instances in which the entire colon is aganglionic is generally the same as with distal colonic aganglionosis. The abdomen is not likely to be as distended and barium enema reveals a microcolon.\(^{(10)}\)

**Other rare presentations have been reported:**

Venugopal, *et al.*, (1997),\(^{(15)}\) they reported, rare presentation of HD as colonic volvulus. HD presenting as sigmoid volvulus in a newborn.

Sarioglu, *et al.*, (1997)\(^{(16)}\) they reported in their study among 302 patients with HD, two patients were admitted with colonic volvulus. Involved segments were caecum and segmoid colon.

Neilson, *et al.*, (1990) they reported delayed presentation of HD: acute obstruction secondary to megacolon with transverse colonic volvulus, despite advances in the diagnosis of HD, occasionally there is still a delay in presentation. They report an unrecognized case of HD in a patient who presented at 11 years of age with volvulus of the transverse colon.\(^{(17)}\)
**Differential diagnosis:**

The differential diagnosis of HD includes hypothyroidism, meconium plug syndrome, colonic neuronal dysplasia, adynamic ileus associated with sepsis, intestinal pseudo-obstruction, if being born to a narcotic addict. These conditions are also associated with delayed passage of meconium at birth.\(^{(9)}\)

Congenital aganglionosis must be distinguished in the neonate from other causes of intestinal obstruction. Later it must be distinguished from acquired megacolon.\(^{(10)}\)

**Diagnosis:**

- **Clinical:**

  The diagnosis of HD is usually not difficult beyond the neonatal period. Constipation with infrequent spontaneous passage of stool dating from early infancy is an important part of history.

  Rectal examination usually reveals an empty ampulla, in extreme instances the abdominal wall is stretched and the veins are prominent, large faecal masses may be palpable over the left colon, in some instances, nutrition is obviously poor.\(^{(10)}\)

- **Radiological:**

  Barium enema X-ray studies usually will confirm the diagnosis because of the characteristic transition from the
narrowed distal rectal or rectosigmoid segment to the more dilated proximal colon. This finding is best demonstrated in a lateral view. It may not be useful in infant with meconium retention, because sufficient time will not have elapsed for the differences in calibre to become prominent. Among patients with acquired megacolon dilatation extends, all the way to the anal canal with no obvious transitional zone.\(^{(10)}\)

- **Rectal biopsy:**

  In doubtful cases, the diagnosis requires a rectal biopsy. The presence of normal numbers of ganglion cell, exclude the diagnosis.

  **(A) Mucosal suction biopsy:**

  Mucosal suction biopsy is satisfactory in many instances. It is the initial procedure of choice, because it is easily performed in Outpatients Department and requires no anaesthesia. The biopsy should include the muscularies mucosae to show the presence or absence of ganglia in Meissner's plexus.\(^{(10)}\)

  **(B) Full thickness rectal biopsy:**

  Absence of ganglia alone, in a specimens obtained by mucosal biopsy does not confirm the diagnosis and should be followed by a full thickness biopsy obtained at least 3 cm proximal to pectinate line. Diminution or absence of ganglin cells distal to
this point is difficult to interpret, since their absence does not establish HD. Careful measurement, proximal to the internal sphincter indicate that myentric ganglia may be absent in normal infant, over a distance of 4 cm, in this segment, none may be seen in the deep submucosal layer for 7 mm and in the superficial submucosal layer for 10 mm.(10)

Ghosh, et al., (1988) in their study about rectal biopsy in the investigation of constipation. A retrospective review of 186 rectal biopsies from 141 children, comparing the age at onset of symptoms with diagnosis of HD. The results of 17 children with HD had the onset of symptoms before the age of 4 weeks, they reported if the age at onset of constipation is after the neonatal period, a rectal biopsy in unnecessary.(18)

- **Manometry:**

Physiologic tests may aid with diagnosis in doubtful cases, especially when the aganglionic segment is short and less easily detectable by X-ray film, short segment HD is also often missed by biopsy. The most important test of pathophysiology is based upon the response of the anal sphincter to distension of the rectum. In contrast to normal individuals and to patients with acquired megacolon, the internal sphincter in patients with HD fails to relax (contract) after distension of the rectum.(10)
• **Histochemistry:**

Histochemistry of a superficial rectal biopsy specimen (staining for acetylcholinesterase) is diagnostic.\(^{(10)}\)

**Treatment:**

At first presentation; management is that of intestinal obstruction, with nasogastric tube aspiration and intravenous fluid and electrolytes replacement, because of the possibility of enterocolitis, antibiotics should be given.\(^{(17)}\)

Once the diagnosis is established definitive surgical operations is the treatment. Preliminary decompression by colostomy may be necessary to relieve obstruction in infants for whom it is decided to postpone definitive surgery, or in older individual, in whom diversion of faecal stream may be necessary to provide a period in which nutritional status can be improved.\(^{(10)}\)

Serial rectal wash out and digital dilatation of the rectum are performed before the pull through is begun. The last of the rectal irrigation has 1% neomycin added to it. Broad spectrum intravenous antibiotic are administered before surgery.\(^{(20)}\)

Hackam, *et al.*, (1997)\(^{(21)}\) reported a series of 109 patients with HD admitted between 1991 and 1996, a definitive diagnosis obtained via rectal biopsy in all patients. One hundred and five of patients were included in the study, 21 patients treated by primary repair without colostomy (SS), 84 patients were treated by colostomy followed by repair (MS). Both SS and MS groups were similar in gender, age, and weight at diagnosis and presence of comorbid illnesses. Reported SS repair represents a safe and effective method in the treatment of HD. The associated complications, including enterocolitis are not significantly different from those associated with MS repair. The hospital and surgical costs of SS repair are significantly reduced compared with MS repair, making this method economically attractive and also eliminate stomal-related complications.\(^{(21)}\)

Luis, *et al.*, in 2000\(^{(22)}\) in their study about. Transanal versus open endorectal pull-through for HD. They reported, transanal endorectal resection and pull-through technique has advantages including eliminating the time taken to open and close the laparotomy and to perform the colectomy followed by pull-through during open procedures, laparotomy is avoided, the colectomy is completed while the pull-through is being performed, and the colonal anastomosis is immediately sutured when the normoganglionic segment is reached.

Marty, *et al.*, in 1995 \(^{(23)}\) they reported in their study of 172 patients, over 22 years. Routine postoperative rectal irrigation by normal saline has significantly decreased the incidence and
severity of enterocolitis in children after surgical correction of HD. The parents were instructed in the irrigation technique before leaving the hospital. Irrigations were started 1 to 2 weeks postoperatively and were performed two times a day for 3 months, then once a day for an additional 3 months. There were no complications from the irrigations themselves.

The surgical management of the condition depends on the clinical presentation. In a baby with obstruction it may be possible to clear the bowel content by gentle rectal irrigation with warm saline; alternatively an emergency colostomy should be performed. In the absence of acute obstruction the diagnosis can be established by investigations and a colostomy then fashioned prior to the definitive surgery. According to preference, either transverse loop colostomy is made or sigmoid loop colostomy using the distal part of normal bowel, the definitive operation follows after. When the condition presents in the newborn, it is customary to delay the procedure until the baby is 9 months or reach 10 kgs in weight. Excision of the aganglionic segment in HD presents two problems: first, the technical difficulties of low anastomosis close to the ano-rectal junction, and second, the risk of damage to the pelvic splanchnic nerves. (24)
Operations:

1. Swenson’s operation (1948) the bowel is divided above the aganglionic segment and the two ends oversewn. The aganglionic bowel is then drawn inside-out through to the perineum by forceps introduced through the anal canal. Normal bowel is drawn down through the invaginated abnormal segment and anastomosed 2 cm from the mucocutaneous junction. The anastomosis is then pushed back through the anal opening to its normal position (24).

Sherman, et al., in 1989 (25) in their study, A 40- years multinational retrospective study of 880 swenson procedures, in seven cities in North American and Western Europe. A follow-up evaluation was obtained on 814 patients, the length of follow-up averaged 10.3 years. The overall postoperative mortality was 2.4% during the entire 40 years of the study. Significant factors influencing postoperative mortality included Down’s syndrome, the patient’s age at the time of the operation, and leak of the distal colonic anastomosis. Most of the patients followed for over 5 years have normal bowel habits, report one to three bowel movements per day, and have no soiling. No patient has urinary incontinence or impotence.
YuZuo, *et al.*, in 2002\(^{(26)}\) they investigate long-term outcome and quality of life after the swenson operation, 45 patients, there were 37 boys and 8 girls, the mean age at the time of operation was 19 months range (2.5 – 60), underwent the Swenson procedure for HD, they were followed-up for 8 to 16 years. Twenty-three patients (51.1\%) had bowel dysfunction. Seventeen patients (37.8\%) suffered from fecal soiling. School absence occurred in 6(13.3\%) patients. Seven patients (15.6\%) had problem in peer relationships.

2. Duhamel’s operation: (1960) normal bowel is divided. A tunnel is made from the pelvic floor posterior and lateral to the rectum and opened into the posterior wall of the anal canal at the level of dentate line. Normal bowel is drawn down through the tunnel so that it lies alongside the rectum. The adjacent walls of the rectum and normal bowel are stapled and divided longitudinally so as to form a common cavity. Rectal sensation is thus preserved within the cavity, the normal bowel provides peristalsis and division of the internal sphincter reduces outlet resistance.\(^{(24)}\)

Martin has modified this procedure by excising the anterior wall of the ganglionic colon and posterior wall of the aganglionic rectum, using stapling device.\(^{(8)}\)
David, et al., in 2002\(^{(27)}\) reported in their study of 29 children with HD in 22 of them the disease found in the first two months of life and in 11 it was noted in the first week of life. In 19 (66.1\%) the aganglionosis restricted to rectosigmoid, underwent one-stage Duhamel’s procedure, with follow-up range (5-7 years). No difference in functional outcome after SS or MS Duhamel-martin procedure. The majority of children seem to fare well with restrictive need of laxatives. The advantage of SS procedure is the prevention of stoma related complication, 1 or 2 additional operation, and extra scar formation.\(^{(27)}\)

Sachiyo, et al., in 1998\(^{(28)}\) studied at Kyushu University Hospital, from 1963 to 1997, 127 patients with HD underwent Z-shaped anastomosis, they reported Z-shaped anastomosis is one of the modification of Duhamel’s procedure that was designed to eliminate the blind rectal pouch and to achieve complete resection of the colorectal septum. It has been the most widely performed operation in Japan for many years. The incidence of soiling in patients who underwent Z-shaped anastomosis was found to be lower than the incidence reported with other procedures. The function of evacuation in these patients improved with age.

3. Soave’s anorectal pull through (1960) the muscle wall of the rectum is left in situ. The mucosa is stripped and the normal
bowel brought through the muscular sleeve, so that the distal dissection to the anal canal can be performed in a sub-mucosal plane. Normal bowel is drawn down through the muscle cuff and prolapsed outside beyond the anal opening. After an interval (usually 3 to 4 weeks) during which the normal bowel becomes fixed in position by adhesions, the stump can be excised. In a modification of this original procedure, which eliminates the need for a second operation, direct anastomoses is done to anal canal. \(^{(24)}\)

4. Myectomy has been used in combination with anterior resection in children with more extensive aganglionosis. (Anorectal myectomy is performed with the child in lithiotomy position. An incision is made at the mucocutaneous junction on the posterior wall of the anal canal and a plane dissected between mucosa and underlying muscle. The strip of muscle, 0.5 to 1.0 cm in width, including the internal sphincter, is excised along the length of the ganglionic segment. \(^{(24)}\)

5. Trans anal mucosal proctectomy with low colo anal anastomosis has been used in adults for the treatment of rectal malignancies, ulcerative colitis and familial polyposis. Trans anal endorectal colonal anastomosis is a good technique for treatment of HD with few operation related complications. \(^{(29)}\)
Some 15% of affected children continue to suffer from bouts of abdominal distension, constipation and enterocolitis, suggestive of a dysfunctional residual colon despite apparent histological normality.\(^{(19)}\)

Blane, et al., in 1994\(^{(30)}\) they reviewed retrospectively the abdominal radiographs in their series to identify specific radiographic characteristic of enterocolitis. A total of 55 episodes of enterocolitis with and abdominal series at presentation were located in the files of 43 patients following pull. Through surgery for HD concludes that the constellation of an intestinal cutoff sign and at least two air-fluid levels on the abdominal series strongly suggests the diagnosis.

Kathleen, et al., in 2002\(^{(31)}\) studied stooling and manometric findings after primary pull-through in HD: perineal versus abdominal approaches. Over 2 years, 26 pull-through procedures were performed. Nine of those were perineal, and 17 were abdominal. Mean follow-up post-pull-through was 23 ± 2.3 months for transabdominal and 14±1.9 months for perineal. Manometric sphincter pressure and enterocolitic episodes after a perineal appear to be similar to results obtained with a conventional transabdominal. This suggests that both methods are
safe, and the perineal does not appear to compromise sphincter integrity.

**Prognosis:**

If enterocolitis is avoided by early diagnosis and treatment, survival is the normal. The quality of life depends on the success of definitive operation and should be near normal in more than 90% of patients.\(^6\) Survival achieved in more than 90% of cases. Long term follow up is important. Most patients (more than 96%) are continent, but soiling is a problem in 2 to 3% of patients. Some patients (10 to 20%) may have constipation, but this can usually be improved with high fiber diet and stool softeners, Most children with postoperative symptoms improve with age. In some children with persistent obstructive symptoms, coexisting intestinal neuronal dysplasia may be a factor. Many of deaths have been observed in babies with Down’s syndrome.\(^9\)

The advances in the management of HD gives most patients a satisfactory outcome. However, some patients continue to have persistent bowel dysfunction including enterocolitis in 6% to 20%, constipation and soiling in 11% to 35%. Some reported the association of HD and intestinal neuronal dysplasia in 25% to 35% and relation to post operative bowel dysfunction.\(^{32}\)
OBJECTIVES

The objectives of this study to describe:

i. The pattern of presentation and methods of diagnosis of HD.

ii. The mode of operative treatment offered to those patients and outcome.

PATIENTS AND METHODS
1. Study design:

This is descriptive study “prospective and retrospective” supported by follow- up of patients, in-patients and outpatients.

2. Study population:

The study population consisted of non-selected patients with a confirm diagnosis of HD; all male and female patients at any age on presentation who has been admitted and offered surgical treatment related to HD.

3. Place and time of the study:

- S.U.H, Department of pediatric surgery.
- The period from January 1999 to March 2003.

4. Data sources:

i- Patients records in the time period specified.
ii- Information obtained by the investigator through examining patients and interviewing parents at the wards and outpatient.
5. Data collection:

Data was collected by a self-administered questionnaire. This was constructed in sections to address the different aspects of the study as follows:

Section  A: Personal data.

B: Presentation.

C: Drug and family history.

D: Abdominal examination findings.

E: Investigations.

F: Surgical management and outcome.

6. Patients assessment:

This was done in the normal manner by history, examination and investigations.

History of personal data, presenting symptoms, drug and family history.

Enterocolitis (preoperative and postoperative) diagnosed in patients with diarrhoea and abdominal distention ± fever, abdominal pain and bleeding per-rectum.

Examination: Concentrate mainly in the abdominal examination, abdominal distension, visible peristalsis, erythema of abdominal wall, palpation for tenderness, any mass and abdominal
organs completed by per-rectal examination findings. Examination for associated anomalies.

7. Investigations:

All patients were evaluated preoperatively by haemoglobin, urea and electrolytes.

Plain abdominal X-ray features of HD looked for are:

- Airless rectum.
- Features of large bowel obstruction:
  - Presence of gas in supine film.
  - Air fluid level in erect film.

Barium enema features of HD include:

- Proximal dilatation with coning into a narrowed segment in the transitional zone.

All patients had rectal biopsy showed absent ganglia, with presence of thick nerve trunk in some biopsies.

No patient underwent manometry or histochemical investigations.

8. Operative management:

All the patients underwent diversion of stool before definitive surgery either by transverse or sigmoid loop colostomy.
Before definitive operation all patients underwent bowel preparation. Prophylactic antibiotic was given and continued into the postoperative period.

The definitive surgical procedure in all patients was Swenson’s procedure either two stages or three stages.

9. Postoperative assessment:

Immediately postoperative for fever, abdominal distension, abdominal pain, diarrhoea and vomiting. Then the status of the wound, leak and abscess. Later on for constipation or stenosis, distension, soiling and impotence and other complications.

10. Review of the literature:

This started before collecting data of this study. Review sources included, medline, journals and textbooks to cover the various aspects.

11. Data analysis:

All the collected data was entered into a master sheet and then analyzed by using Statistical Package of Social Sciences (SPSS), the result expressed in numbers and percentage.
RESULTS

From January 1999 to March 2003 a total of 40 patients were admitted and treated at S.U.H, with confirmed diagnosis of Hirschsprung’s disease (HD).

- Thirty-five patients (87.5%) were males and 5 patients (12.5%) were females, male to female ratio 7:1 (Fig. 1).
- The age of the patients on presentation ranging from 3 days to 16 years, with mean of $39.03 \pm 47.11$ months (Fig. 2).
- The 40 patients were resided in different regions in the Sudan (Table 1).
- In 12 patients (30%) their parents were first degree relatives (Table 2).
- Positive family history of HD in 3 patients (7.5%) (Fig. 3).
- Associated anomalies were found in 4 patients (10%). (Table 3).
- Twenty-eight patients (70%) had a history of enema or laxative use before diagnosis (Fig. 4).
- The aganglionic segment confined to the rectum in 17 patients (42.5%) (Table 4).
Symptoms in the study group (Fig. 5):

- Failure to pass meconium in the first 24 hours of life was encountered in 36 patients (90%), constipation in 36 patients (90%) and abdominal distension in 38 patients (95%).
- Vomiting was reported in 21 patients (52.5%).
- Three patients (7.5%) presented with enterocolitis.

Abdominal examination findings on presentation:

- **Abdomen was distended in 36 patients (90%)** with visible peristalsis in 10 (25%) (Table 5).

- Abdomen was soft in 38 patients (95%) (Fig. 6)

- Per-rectal examination revealed empty rectum in 27 patients (67.5%) with gush of gas or faces in 16 patients (40%).

Radiological findings:

- Plain abdominal x-ray showed features of HD in 27 patients (67.5%) and barium enema in 13 patients (32.5%).

Surgical treatment:

- All patients underwent colostomy at the age corresponding to the age of presentation (Fig 7).
• All patients underwent swenson’s procedure as definitive surgery (Fig. 8).

• The age of definitive surgery was ranging from one year to 16.5 years, with mean of 4.07 ± 3.8 years (Fig. 9).

Operative outcome:

• *Postoperative complications occurred in 13 patients (32.5%) (Fig. 10).*

• *Postoperative enterocolitis occurred in 5 patients (12.5%).*

• *Bowel obstruction occurred early in the first days following surgery in 6 patients (15%) (Table 6).*

• Three patients (7.5%) had postoperative enterocolitis and bowel obstruction (Table 7).

• Two patients (5%) had postoperative enterocolitis and anastomotic stricture (Table 8).

• Wound infection occurred in 7 patients (17.5%).

• Stool incontinence (soiling) occurred in 2 patients (5%).

Mortality:
Two patients (5%) died postoperatively one male and the other female, one had Down’s syndrome and both developed postoperative enterocolitis.

**Table 1: Distribution of patients according to their residence**

<table>
<thead>
<tr>
<th>Region</th>
<th>Frequency</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Northern</td>
<td>14</td>
<td>35.0%</td>
</tr>
<tr>
<td>Capital</td>
<td>9</td>
<td>22.5%</td>
</tr>
<tr>
<td>Centre (Gezira)</td>
<td>8</td>
<td>20.0%</td>
</tr>
<tr>
<td>Western</td>
<td>5</td>
<td>12.5%</td>
</tr>
<tr>
<td>Eastern</td>
<td>3</td>
<td>7.5%</td>
</tr>
<tr>
<td>Southern</td>
<td>1</td>
<td>2.5%</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>40</strong></td>
<td><strong>100%</strong></td>
</tr>
</tbody>
</table>

**Table 2: Demonstrates parents consanguinity**

<table>
<thead>
<tr>
<th>Consanguinity</th>
<th>Frequency</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>First degree cousins</td>
<td>12</td>
<td>30%</td>
</tr>
<tr>
<td>Second degree cousins</td>
<td>4</td>
<td>10%</td>
</tr>
<tr>
<td>Not relatives</td>
<td>24</td>
<td>60%</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>40</strong></td>
<td><strong>100%</strong></td>
</tr>
</tbody>
</table>
Table 3: Associated anomalies

<table>
<thead>
<tr>
<th>Anomalies</th>
<th>Frequency</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>No associated anomalies</td>
<td>36</td>
<td>90%</td>
</tr>
<tr>
<td>U.D.T</td>
<td>2</td>
<td>5%</td>
</tr>
<tr>
<td>Down’s syndrome</td>
<td>1</td>
<td>2.5%</td>
</tr>
<tr>
<td>V.S.D</td>
<td>1</td>
<td>2.5%</td>
</tr>
<tr>
<td>Total</td>
<td>40</td>
<td>100%</td>
</tr>
</tbody>
</table>

Table 4: Illustration of level of aganglionic segment after surgery

<table>
<thead>
<tr>
<th>Level</th>
<th>Frequency</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rectum</td>
<td>17</td>
<td>42.5%</td>
</tr>
<tr>
<td>Rectosigmoid</td>
<td>21</td>
<td>52.5%</td>
</tr>
<tr>
<td>Descending colon</td>
<td>2</td>
<td>5%</td>
</tr>
<tr>
<td>Status of the abdomen</td>
<td>Frequency</td>
<td>Percentage</td>
</tr>
<tr>
<td>-------------------------------------------</td>
<td>-----------</td>
<td>------------</td>
</tr>
<tr>
<td>Distended with visible peristalsis</td>
<td>10</td>
<td>25%</td>
</tr>
<tr>
<td>Distended without visible peristalsis</td>
<td>26</td>
<td>65%</td>
</tr>
<tr>
<td>Not distended</td>
<td>4</td>
<td>10%</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>40</strong></td>
<td><strong>100%</strong></td>
</tr>
</tbody>
</table>
Table 6: Correlation between site of colostomy and bowel obstruction

<table>
<thead>
<tr>
<th></th>
<th>Bowel obstruction</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>Colostomy sigmoid</td>
<td></td>
<td></td>
</tr>
<tr>
<td>count</td>
<td></td>
<td></td>
</tr>
<tr>
<td>% within colostomy</td>
<td>6</td>
<td>27</td>
</tr>
<tr>
<td>% within bowel obstruction</td>
<td>18.2</td>
<td>81.8%</td>
</tr>
<tr>
<td></td>
<td>100%</td>
<td>79.4%</td>
</tr>
<tr>
<td>Transverse</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Count</td>
<td></td>
<td></td>
</tr>
<tr>
<td>% within colostomy</td>
<td>7</td>
<td>5</td>
</tr>
<tr>
<td>% within bowel obstruction</td>
<td>100%</td>
<td>100%</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Count</td>
<td>6</td>
<td>34</td>
</tr>
<tr>
<td>% within colostomy</td>
<td>15%</td>
<td>85%</td>
</tr>
<tr>
<td>% within bowel obstruction</td>
<td>100%</td>
<td>100%</td>
</tr>
</tbody>
</table>

P = 0.473
Table 7: Correlation between postoperative enterocolitis and bowel obstruction

<table>
<thead>
<tr>
<th>Enterocolitis</th>
<th>Yes count</th>
<th>Bowel obstruction</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>% within enterocolitis</td>
<td>60%</td>
<td>40%</td>
<td>100%</td>
</tr>
<tr>
<td>% within bowel obstruction</td>
<td>50%</td>
<td>5.9%</td>
<td>12.5%</td>
</tr>
<tr>
<td>No</td>
<td>Count</td>
<td>3</td>
<td>32</td>
</tr>
<tr>
<td>% within enterocolitis</td>
<td>8.6%</td>
<td>91.4%</td>
<td>100%</td>
</tr>
<tr>
<td>% within bowel obstruction</td>
<td>50%</td>
<td>94.1%</td>
<td>87.5%</td>
</tr>
<tr>
<td>Total</td>
<td>Count</td>
<td>6</td>
<td>34</td>
</tr>
<tr>
<td>% within enterocolitis</td>
<td>15%</td>
<td>85%</td>
<td>100%</td>
</tr>
<tr>
<td>% within bowel obstruction</td>
<td>100%</td>
<td>100%</td>
<td>100%</td>
</tr>
</tbody>
</table>

P = 0.003
Table 8: Correlation between postoperative enterocolitis and anastomotic stricture

<table>
<thead>
<tr>
<th></th>
<th>Anastomotic stricture</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>Enterocolitis</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>% within enterocolitis</td>
<td>40%</td>
<td>60%</td>
</tr>
<tr>
<td>% within anastomotic stricture</td>
<td>33.3%</td>
<td>9.7%</td>
</tr>
<tr>
<td>No</td>
<td>4</td>
<td>31</td>
</tr>
<tr>
<td>% within enterocolitis</td>
<td>11.4%</td>
<td>88.6%</td>
</tr>
<tr>
<td>% within anastomotic stricture</td>
<td>66.7%</td>
<td>90.3%</td>
</tr>
<tr>
<td>Total</td>
<td>6</td>
<td>34</td>
</tr>
<tr>
<td>% within enterocolitis</td>
<td>15%</td>
<td>85%</td>
</tr>
<tr>
<td>% within anastomotic stricture</td>
<td>100%</td>
<td>100%</td>
</tr>
</tbody>
</table>

P = 0.043
Fig. 1: Sex distribution in the study group

Male 35 (87.5%)
Female 5 (12.5%)

Fig. 2: Patients distribution according to age on presentation

<table>
<thead>
<tr>
<th>Age group (in months)</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-1</td>
<td>4 (10%)</td>
</tr>
<tr>
<td>2-12</td>
<td>13 (32.5%)</td>
</tr>
<tr>
<td>13-60</td>
<td>14 (35%)</td>
</tr>
<tr>
<td>&gt; 60</td>
<td>9 (22.5%)</td>
</tr>
</tbody>
</table>
Fig. 4: Summary of drug history

<table>
<thead>
<tr>
<th>Drug used</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Laxatives or enema</td>
<td>70%</td>
</tr>
<tr>
<td>Antibiotics</td>
<td>30%</td>
</tr>
<tr>
<td>Other</td>
<td>10%</td>
</tr>
</tbody>
</table>

Fig. 5: Illustration of symptoms in the study group

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Failure to pass meconium</td>
<td>90%</td>
</tr>
<tr>
<td>Constipation</td>
<td>90%</td>
</tr>
<tr>
<td>Abdominal distension</td>
<td>95%</td>
</tr>
<tr>
<td>Vomiting</td>
<td>52.5%</td>
</tr>
<tr>
<td>Abdominal pain</td>
<td>20%</td>
</tr>
<tr>
<td>Fever</td>
<td>17.5%</td>
</tr>
<tr>
<td>Diarrhoea</td>
<td>10%</td>
</tr>
<tr>
<td>Failure to thrive</td>
<td>10%</td>
</tr>
</tbody>
</table>
Fig. 6: Illustration of abdominal status on palpation

- Soft abdomen: 38 (95%)
- Tender abdomen: 2 (5%)

Fig. 7: Site of loop colostomy

- Sigmoid loop colostomy: 33 (82.5%)
- Transverse loop colostomy: 7 (17.5%)
Fig. 8: Stages of Swenson's procedure

- Two stages: 33 (82.5%)
- Three stages: 7 (17.5%)
Fig. 9: Patients distribution according to age of definitive surgery
Fig. 10: Illustration of postoperative complications

- Enterocolitis: 5(12.50%)
- Wound infection: 6(15%)
- Bowel obstruction: 6(15%)
- Constipation: 7(17.50%)
- Leak: 2(5%)
- Soiling: 2(5%)
- Anastomotic stricture: 2(5%)
DISCUSSION

This is a descriptive retrospective and prospective study conducted at SUH, the main referral hospital for paediatric surgical cases in Sudan. Forty patients with HD were treated surgically by Swenson's pull through procedure. Male to female ratio 7:1 (35 males and 5 females), not in accordance with the reported
literature, Ellis\(^{(6)}\) reported male to female ratio was 4: 1. Yuzuo\(^{(26)}\) reported male to female ratio was 4.5: 1. Generally males were the most to be affected and our series went parallel to this, however, the ratio was higher than the reported literature.

The residence of 23 patients (57.5\%) of our series from states away from the centre, 14(35\%) are coming from the North, 5(12.5\%) from the West, 3(7.5\%) from the east and one patient (2.5\%) from the South, the other 17 patients (42.5\%) from the capital and centre of Sudan.

In our series 70\% of patients, had a history of laxtive or enema use, 30\% were using an antibiotics and 10\% are using other modalities, such as soap, Helba and cauterization.

The mean age at presentation was \(39.03 \pm 47.11\) months, ranging from 3 days to 16 years (reflect late presentation of our patients), neonatal presentation in 4 patients (10\%), infants in 13 patients (32.5\%) and 23(57.5\%) presented above one year of age. Sleisenger\(^{(10)}\) stated HD is a neonatal disease. David\(^{(27)}\) reported in the study of 29 patients in 22 patients (76\%), the disease presented in first two months of life. The late presentation in our study group can be explained by far residence of most of patients from specialized hospitals and unawareness of treating medical
personnel about HD reflected use of laxatives, enemas and antibiotics.

HD is a familial disease in our series, 3 patients (7.5%) had a positive family history and 12 patients (30%) their parents were first degree relatives. Sleisenger\(^{(10)}\) reported in a study of 326 patients, the incidence among siblings was 3.6% and consanguinity is exceptional, only three instances have been reported. In our study group there is a high familial index. This can be explained by the high incidence of consanguinity in our society. Association of HD with Down's syndrome was reported with increase of risk of complications,\(^{(6,10,13,25)}\) approximately 2% of patients with HD have Down's syndrome,\(^{(10)}\) in our series 2.5% of patients had Down's syndrome more or less similar to other series. Other associated anomalies reported include: megacystis and megaureter, hydrocephalus, V.S.D, cystic deformities of the kidney, U.D.T, diverticulum of the urinary bladder, imperforate anus, Meckle's diverticulum, hypoplastic uterus, polyposis of the colon, ependymoma of the fourth ventricle and Laurence-Moon-Biedl-Bardet syndrome.\(^{(10)}\) In our series 2 patients (5%) had U.D.T and one patient (2.5%) had V.S.D, the other anomalies were not reported in our series, this could be explained by the series being small.
The aganglionic segment in our series was confined to the rectum in 17 patients (42.5%), rectosigmoid in 21(52.5%) and descending colon in 2(5%). The rectum and rectosigmoid junction are common sites for aganglionic segment.\(^{(9,10,11)}\) Jennifer\(^{(12)}\) mentioned, the greatest distribution of aganglionic segment in the rectum (30%) and rectosigmoid (44%), our findings approximately in accordance. In our series there is no case of total colonic aganglionosis reported.

In our series the main symptoms were delayed passage of meconium in the first 24 hours of life (90%), constipation (90%) - started in the neonatal period- and abdominal distension (95%), this goes with the literature.\(^{(9,10)}\) Other symptoms include: vomiting 21(52.5%), abdominal pain 8(20%), fever 7(17.5%) and failure to thrive in 4 patients (10%). Jennifer\(^{(12)}\) described the typical presentation of HD consists of failure to pass meconium spontaneously within the first 24 hours of life followed by constipation, abdominal distension, bilious vomiting and failure to thrive if not detected in the newborn period.

In our series, 3 patients (7.5%) had preoperative enterocolitis. Postoperative enterocolitis not related to any patient with preoperative enterocolitis. Elhalaby\(^{(14)}\) reported in a series on 168 patients with HD, preoperative enterocolitis occurred in
13(7.7%) and characterized by abdominal distension and explosive diarrhoea associated with the intestinal cutoff sign. Sleisenger\textsuperscript{(10)} also reported repeated enemas, which are followed by enterocolitis, our findings goes with Elhalaby and Sleisenger statement.

In our study group the abdomen was found distended in 36(90%) with visible peristalsis in 10(25%) and soft in 38(95%). Per rectal examination revealed empty rectum in 27(67.5%) with gush of gas and faeces in 16(40%), this goes with the literature.\textsuperscript{(7,8,10)}

Radiological finding in our patients, plain abdominal x-ray showed features of HD in 27(67.5%) and barium enema was diagnostic in 13(32.5%), those were a total group of patients subjected to radiology. Jennifer\textsuperscript{(12)} stated, barium enema may reveal the sharp contrast in size between the dilated normal colon and the aganglionic bowel. Sleisenger\textsuperscript{(10)} reported, neither plain X-ray abdomen nor barium enema is diagnostic. In our series although all the subjected patients for radiology showed features of HD, you can not depend on radiology because there was no comparison group with negative tissue diagnosis and positive radiology, therefore, a tissue diagnosis is a must for diagnosis.
All patients, in our series were offered definitive surgical treatment in form of Swenson's procedure either two stages in 33(82.5%) or three stages in 7 patients (17.5%). Stomal related complication observed in two patients, one had a prolapse of bowel which was reduced then followed by surgery, the other had colostomy stenosis which was revised. The age of definitive surgery ranges from 7 months to 16.5 years, with a mean of 4.08 ± 3.8 years. After definitive surgery, postoperative complications occurred in 13 patients (32.5%). Enterocolitis occurred in 5 patients (12.5%), one had Down's syndrome. Bowel obstruction occurred in 3 patients with enterocolitis (P= 0.003) and anastomotic stricture occurred in 2 patients (P= 0.043). Two patients with postoperative enterocolitis died, one had Down's syndrome. The occurrence of explosive diarrhoea in any patient with HD is suggestive of Hischsprung associated enterocolitis with an incidence of 21.4% postoperatively, significant factors for enterocolitis were delay in a diagnosis and the presence of Down's syndrome with incidence of 24% postoperatively, our finding in the study group was less than the reported incidence in the literature. This can be attributed to the small size of our studied group. Postoperative bowel obstruction occurred in 6 patients (15%) in our series, although statistically not significant, all
these patients underwent two stages Swenson's procedure (P= 0.473), and were offered a transverse colostomy. Redo pull-through was performed in three patients of whom two patients had a leak, other 3 patients of this group were treated by a serial rectal dilatation.

In our study group survival was 95%, and stool incontinence occurred in two patients (5%). Survival rate was more than 90%, more than 96% were continent, soiling occurred in 2 to 3%.\(^{(9)}\) Yuzuo\(^{(26)}\) studied 45 patients underwent Swenson's procedure, the mean age was 19 months, 51.1% had bowel dysfunction include enterocolitis in 6 to 20%, constipation and soiling in 11 to 35%, were related to association with intestinal neuronal dysplasia.\(^{(32)}\) Although reported literature have a different percentage about bowel dysfunction, our findings in some with context in other were best inspite of low facilities.

Wound infection occurred in 7 patients (17.5%) with one burst abdomen, all were treated according to the result of culture and sensitivity. In three patients the culture revealed \textit{E. Coli}, in two patients \textit{Pseudomonas} and \textit{Staph. aureus} in two patients. Wound infection in large bowel surgery and the inadequate bowel preparation in some cases "operative findings" can be accepted in our study group.
Postoperative mortality in a study of 880 Swenson's procedure was (2.4%) and factors influencing mortality Down's syndrome, the patient's age at the time of the operation and leak at the distal colonic anastomosis,\(^{(25)}\) many of deaths had been observed in babies with Down's syndrome.\(^{(9)}\) In our series two patients (5%) died, the first 17 months old male, known case of Down's syndrome, presented when he was four month age, immediate diversion of stool through a sigmoid loop colostomy, underwent definitive surgical procedure when he was 17 months, developed postoperative enterocolitis and died after 7 days of operation. The second patient a female, 5 years age, presented when she was 3 years old, sigmoid loop colostomy done for her at that time then followed by Swenson's procedure at the age of 5 years, developed postoperative enterocolitis, septicaemia and death. The main cause of death in our study group was enterocolitis.

**CONCLUSION**

- Failure of spontaneous passage of meconium in the first 24 hours of life is pathognomic symptom of the disease.
• Late presentation of the patients in our series.
• Rectal biopsy is essential for diagnosis.
• Enterocolitis pre or postoperative is a major problem with HD and contributed greatly to mortality.
• Swenson's procedure was the only procedure offered for patients in our study group with good results.
• High failure rate of the patients to attend follow up regularly.

RECOMMENDATIONS

In the light of the data we obtained from this study and also from the literature we draw the following recommendations:

• Increase awareness about the disease e.g. in Medical Schools.
• Early referral to specialized centres, especially in patients with delayed passage of meconium.

• Because of the rarity of the condition and difficulty of prospective study we recommend improving patients records for retrospective data.

• Neonatal surgical units and neonatal intensive care units.

REFERENCES


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Questionnaire
Hirschprung’s disease; Pattern of presentation,
Mode of management and outcome

(A) Personal Data:


(B) Presentation:
1. Failure to pass meconium in first 24 hrs of life  Yes ( ) No ( )
2. Constipation:  Yes ( ) No ( )
3. Abd. Distension:  Yes ( ) No ( )
4. Vomiting:  Yes ( ) No ( )
5. Abd. Pain:  Yes ( ) No ( )
6. Failure to thrive:  Yes ( ) No ( )
7. Diarrhea:  Yes ( ) No ( )
8. Fever:  Yes ( ) No ( )
9. Bleeding per rectum:  Yes ( ) No ( )
10. Other ..........................................................................................................................

(C) Drug History:
1. Use of laxative (which)

...........................................................

2. Use of antibiotic (which) ....................... 3. Other

...........................................................

Family History:
1. Parents

consanguinity........................................

2. Family history of similar condition ...................

D. Abdominal examination:

a. Distended:  Yes ( ) No ( )

If “Yes”, Mild ( ) Moderate ( ) Server ( )

b. Visible peristalsis:  Yes ( ) No ( )

c. Erythema of Abd. Wall:  Yes ( ) No ( )

d. Abdominal palpation  Soft ( ) Tender ( )

e. Abdominal organ: .................................................................
f. Per rectal examination: Rectum empty ( ) Full ( )

Gush of gas and meconium or feces: Yes ( ) No ( )
Associated anomalies: ..............................................................

(E) Investigations:
1. HB gm/dl ......................... 2. Urea & electrolytes.........................
3. Plain abdominal X-ray: Airless rectum: Yes ( ) No ( )
   Gas in the large bowel & dilation: Yes ( ) No ( )
   Fluid level: Yes ( ) No ( )
   Other ...........................................................

4. Barium enema ...........................................................................
   .............................................................................
5. Manometry .............................................................................
6. Rectal biopsy ...........................................................................
7. Other .....................................................................................

(F) Surgical management and complications:
1. Colostomy: Site ________ Age ________ Complications ________
2. Definitive surgery: which procedure:
   Swenson’s ( ) Duhamel’s ( ) Soav’s ( ) Age ............... 
3. Post-operative complications:
   a. Enterocolitis ( ) b. Wound infection ( ) If Yes: culture.........
   c. Bowel obstruction ( ) d. Leak ( ) e. Abscess ( )
   f. Stenosis ( ) g. Soiling ( ) h. Urinary incontinence ( )
   i. Other: ................................................................................