HIRSCHPRUNG'S DISEASE AND OTHER ALLIED DISORDERS AS SEEN AT THE KENYATTA NATIONAL HOSPITAL DURING THE LAST 5 YEARS - (1973-1978) (REVIEW AND ANALYSIS)

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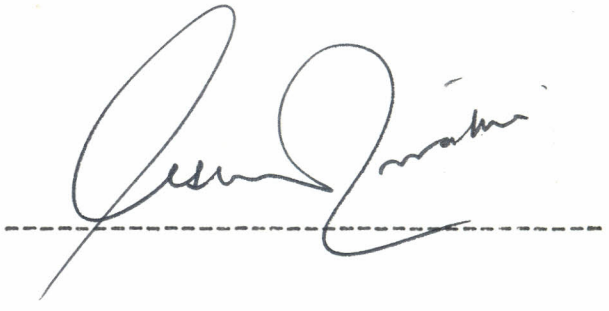
A thesis submitted in part fulfillment for the degree of Master of Medicine (Surgery) 1978.

This thesis is my original work and has not been presented for a degree in any other University.



This thesis has been submitted for examination with my approval.

(Supervisor)

P R E F A C E:

In Africa, the literature on Hirschprung's disease or congenital aganglionosis is very scarce as compared with the literature on this disease in Europe and America. Like many other diseases, it has been even considered a rarity among the Africans (South African Bantus Katz 1966).

In Eastern African countries there is documentation of Hirschprung's disease in Uganda (Shepherd 1966) where the apparent rarity of the disease was emphasized. From Uganda there also is documentation of the non-congenital form of megacolon whose surgical importance as a cause of intestinal obstruction has been shown. (DeSouza 1966).

In Kenya there is, so far, no published document to show the importance of Hirschprung's disease or any of the other allied conditions of the colon.

This dissertation is meant to fill in the gap, though in a localised area of Kenya, and it may, in future, form a starting point for a country wide study of these surgically important diseases.

The dissertation reviews and analyses all cases, in retrospective and prospective study, seen at The Kenyatta National Hospital which is the National Reference Hospital for Kenya; over a period of over five years. (1973 - March 1978).

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INTRODUCTION AND REVIEW OF LITERATURE.

Hirschprung's disease to-day goes by many names as the disease has been understood a step better than it was ninety years ago when Hirschprung wrote on it; "Congenital megacolon," "congenital intestinal aganglionosis" are some of these names. However, Hirschprung's disease as an eponym, seems not only to have withstood the test of times but also to have brought in less confusion on terminologies on the disease. In this paper Hirschprung's disease will be used to mean the same as intestinal aganglionosis as it is understood worldwide at present. Congenital megacolon tends to limit one to only those cases where the aganglionosis is found in a 'short' segment or''Long' segment of the distal colon or rectum leading to dilated colon proximally, but it is known that cases exist whereby the aganglionosis extends to include even the ileum and hence no megacolon.

It has been claimed in literature that before (1886) when Hirschprung presented his paper on congenital megacolon, more than twenty papers had been written on the subject. Notable among these was that by a Dutch Surgeon Frederick Ruyschii who wrote it in Latin in the 17th century.

However, it was to Hirschprung (1888), whose classical treatise on a disease which was later named after him that the credit went. His paper referred to his clinical experience and autopsy findings on a congenital disease which was characterized by high mortality in childhood and at autopsy, a dilated and hypertrophic colon was a notable feature. He also said there was a distal narrow segment of the colon. He focussed his attention to the dilated segment which he claimed was the primary lesion. For many years people thought the primary disease was in the hypertrophic dilated segment, and the pathologic importance of the apparent narrow segment was not appreciated for some years.

Suggestion of the neurogenic nature of megacolon was made by Tittel 1901. He claimed he had noticed some abnormality in the intramural ganglion cells of the colon, in these cases. The same trend of thought was taken by Hawkins (1907) who argued that without a demonstrable mechanical obstruction in the colon, the disease was due to neuromuscular defect:

The inflammatory nature of megacolon was claimed by Cameron (1927). He said he had detected some inflammatory cellular destruction of the intermuscular ganglion cells of the colon.

Etzel(1937) in his series of 8 cases described absence of plexuses in the total distal colonic wall. Three years later, Alvarez (1940) reasonably argued that what Hirschprung had called primary lesion (Hypertophy and dilatation) of the colon was merely a secondary manifestati of loss of function by the ganglion cells in the distal colon.

In the late Forties most of the present logical understanding of intestinal ganglionoses was worked out. Zuelzer (1948); Bodian et al (1949) Whitehouse et al (1948) put to light the principles guiding the pathophysiologic and histopathologic understanding of Hirschprung's disease: In their series of cases they consistently demonstrated absence of ganglion cells of the Auerbach's and Meissner's plexuses and also hypertrophy of the nonmyelinated nerve fibres . These are the basis of present day modes of diagnos and treatment.

The surgical management of cases of Hirschprung's disease was pioneered (Documented) by surgeons like Maunsell (1892) and Treves (1898). The latter had reasoned out that the colonic dilatation was secondary to spasm of the narrow segment and so he resected it out and anastomosed the ends. (His patient was seen 67 years later; Daintree 1957).

The surgical principles of the operation done for cases of Hirschprung's disease were founded by Swenson and Bill (1948). Resection of the aganglionic segment with preservation of the anal sphincter was their

proposition and to-day many modifications of these have been formulated to suit the clinical and histopathologic findings.

To-day the treatment for Hirschprung's disease has been perfected but it is felt by many that as far as aetiology is concerned we are still in the realm of speculations - hence animal studies are being carried out to try and elucidate on this problem of the real cause of aganglionosis. With chicken embryos Yutema (1954), Human featuses of different ages (Okamoto et al 1967) and with mice - (Piebald lethal mutation) Lane (1966)-all efforts are being done to show the cause why either the ganglion cells do not migrate to the aganglionic segment or if they do, why they do not develop normally.

This dissertation comprises of retrospective review and analysis of those cases seen at the Kenyatta National Hospital, referred or seen as new cases, over the period (1973 to March 1978).

The analysis and review of these cases is meant to show that Hirschprung's disease is not as rare as it has been mentioned in some parts of Africa (Katz 1966). Though the work was done at the National Reference Hospital and therefore dealt with selected cases from the whole of Kenya the reflection on the tribal distribution which is analysed may show the national importance of Hirschprung's disease and the other related diseases.

In the dissertation an attempt is made to analyse certain aspects of these diseases which are also compared with some aspects as seen in Europe and America where much time has been studied on these diseases. The ages of the patients at the time of presentation and the time the Symptoms are thought to be of significance by parents or guardians to require medical consultation are reviewed. This aspect is important as Hirschprung's

disease, being a congenital disease, the symptoms start early in life - neonatal period. Any factor which may delay early medical attention - either in failure on the part of parents to appreciate the significance of the symptoms, or, also failure on the part of clinicians - to appreciate the significance of symptoms and signs early - contributes to the mortality in cases of Hirschprung's disease early in life. The review of symptoms and signs and their importance in diagnoses of the cases is compared to what is considered to be the important signs and symptoms where large series have been reviewed in Europe and America. Where other symptoms and signs pertinent to the reviewed cases here have been found, emphasis has been put for the benefit of diagnosis in our conditions.

The tribal distribution has been reviewed with prior understanding that any dispar ity may reflect not on the rarity of the disease among one tribe or another, but on the geographic distribution of these tribes in relation to the Kenyatta National Hospital.

The methods of diagnosis in all cases have been analysed and the management of these cases as well. In Kenya the medical care distribution is limited by lack of trained manpower to diagnose and manage disease appropriately. Besides the scarcity of manpower , diagnostic tools are also scarce. It is with this understanding that I have reviewed the role of the available means of diagnosing Hirschprung's disease as well as other related disorders in order to emphasize which tools could be used effectively in diagnosis. With so many forms of surgical management of Hirschprung's disease being documented in Europe and America, not as many are easy to administer. The simplicity of a surgical operation may not mean effectiveness in curing a disease, but assessing all these forms of operations in light of the available data on the disease may give better planning for training personnel on the better operations within available means. It is attempted here to review all the available data and to compare the methods used in treating surgically the cases of Hirschprung's disease and those

related diseases. The technically possible and effective methods that are practised here have been reviewed in literature and briefly discussed.

The morbidity due to Hirschprung's disease and also those other related disease is analysed and the management and non-management of these condition compared.

I have attempted to show that though Hirschprung's disease is believed to be uncommon, the rarity of the disease is apparent and that by better understanding of the symptoms and signs by the populace, and therefore early presentation to hospital and the same understanding by the clinicians will wipe out the notion of the rarity of Hirschprung's disease.

Similarly I have tried to show the existence of other obscure conditions whose clinical presentations are similar to Hirschprung's disease and they must be excluded in the diagnosis of intestinal aganglionosis as their treatment is not necessarily the same.

MATERIALS AND METHODS:

The source of materials for this analysis of cases of Hirschrung's disease and the allied conditions was from The Kenyatta National Hospital. The period of study of these case records was from 1973 to 1978 (March) - 5 years approximately. The study included all those cases who were diagnosed as Hirschprung's disease or megacolon by any of the existing methods. For the year 1977 and part of 1978 all cases seen in the Hospital and suspected of Megacolon were interviewed (or relatives) personally and investigation done in collaboration with the consultants in charge of the patients. The study, therefore, encompasses retrospective as well as prospective analysis. Where biopsies of the patients (rectal or colonic) were taken the report and processing for staining the slides, collaboration of the Hospitals' Department of pathology was requested. Haematoxylin and Eosin were used to stain and as a separate study, some of the specimens were frozen with "dry ice" for histochemical studies.

All slides were examined with light microscope by the pathologists in charge.

In the analysis, Sex, tribal distribution, the presenting symptoms, age distribution at presentation, time of onset of symptoms; methods of investigations The diagnosis and forms of treatment were studied and presented in tabular form.

RESULTS

Table III (a)

Age in years at presentation and number of cases.

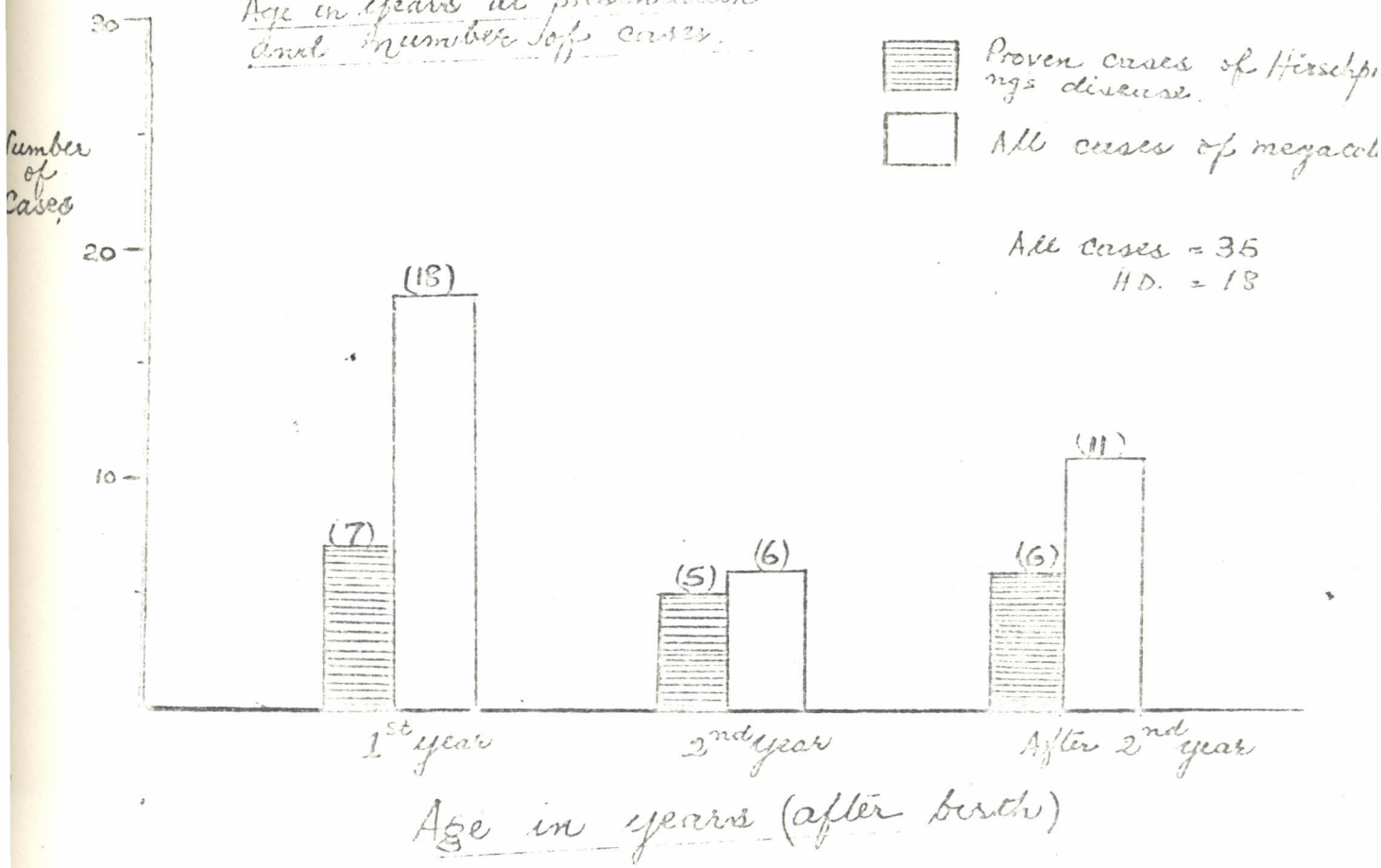
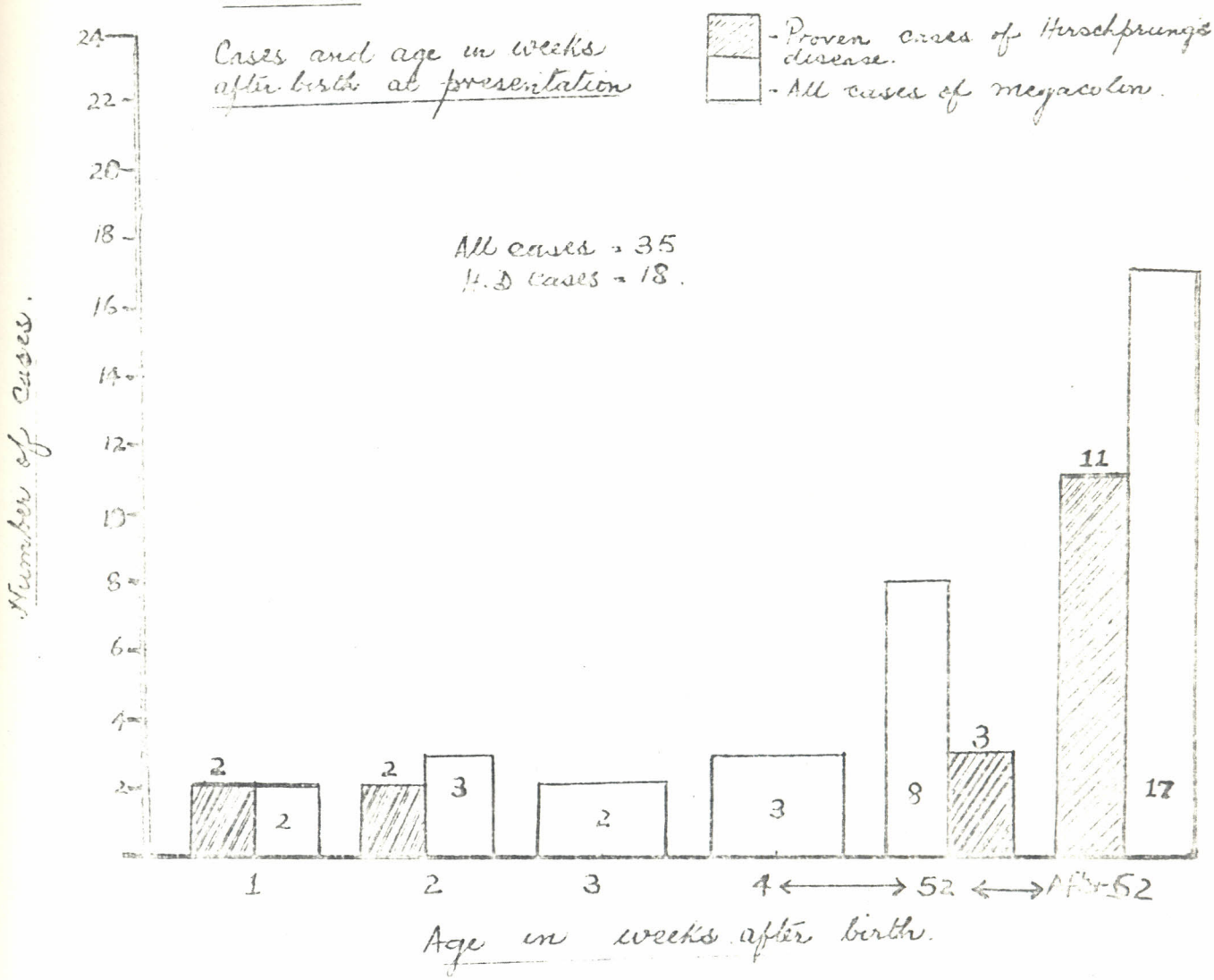


Table III(b)

Cases and age in weeks after birth at presentation



Sex Distribution. Table 1

Sex	No of Cases (all)	%	H.D.	%age
Males	26	74.3%	16	88.9%
Females	9	25.7%	2	11.1%
Total	35	100%	18	100%

(H.D. = Hirschprung's Disease)

Tribal Breakdown of cases.

Table 11

Tribe	All cases	H.D. Cases		Total
		M	F	
Kikuyu	19	10	1	11
Kamba	8	4	0	4
Luhya	3	0	1	1
Luo	4	1	0	1
Taita	1	1	0	1
Total	35	16	2	18

Table IV.

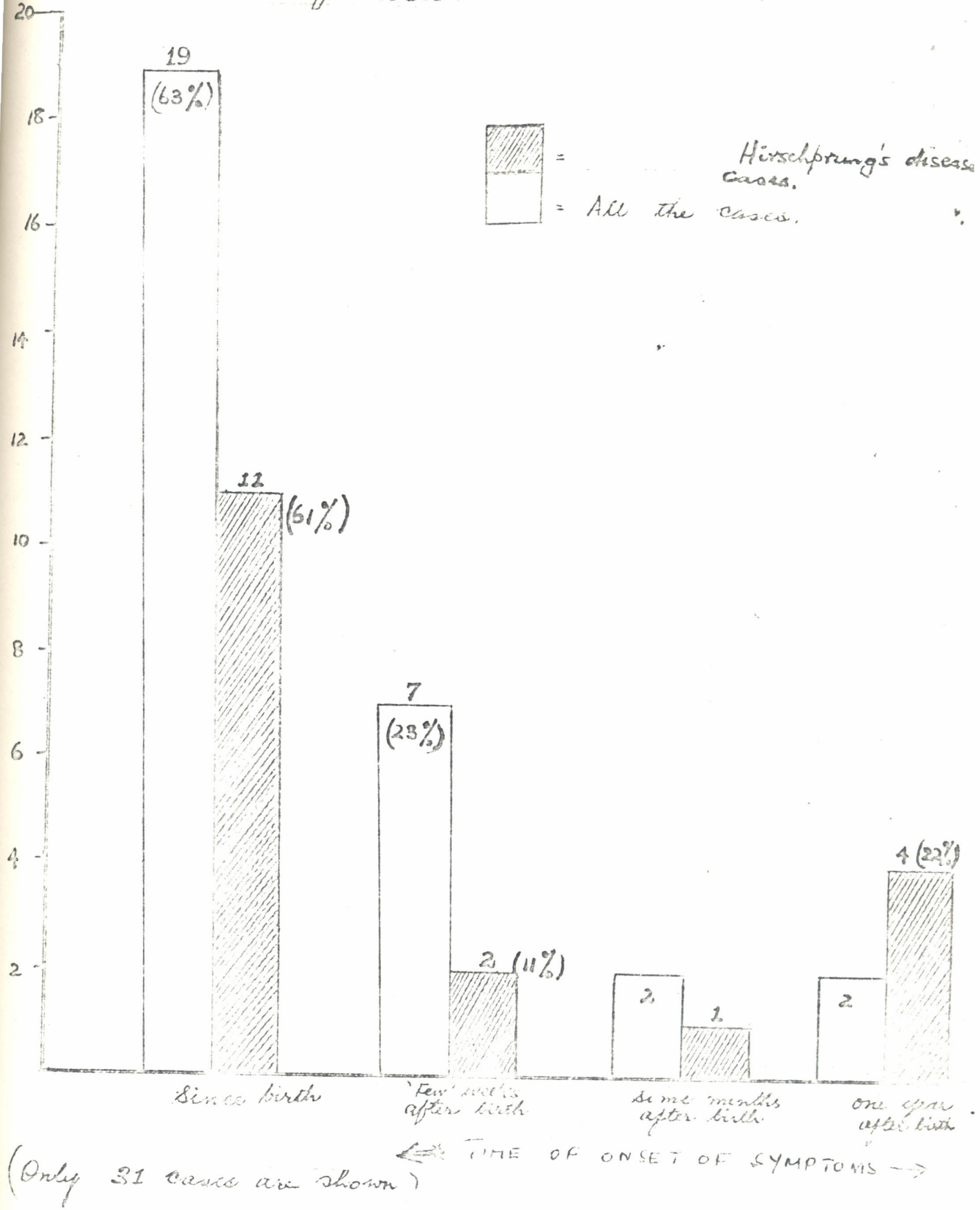
The commonest symptoms of presentation.

Symptoms	All cases	%	H.D	%	%*
(a) Abdominal distension	27	77.1	12	63.0	92%
(b) Constipation	25	71.4	13	69.4	84%
(c) Vomiting	14	40.0	6	35.4	76%
(d) Diarrhea	8	20.0	2	11.7	-
(e) Failure thrive	5	14.3	2	-	-
(f) Others	5	14.3	1	-	-
(g) Combined (a+b+c)	8	23.0	3	-	53%
(h) Combined (a+b)	18	51.4	9	-	-

* In this column the percentages of the frequency of the symptoms are from a series by Daintree on cases of Hirschprung's disease (Daintree 1957)

Table V

Table showing time of onset of Symptoms against No of cases



(Only 31 cases are shown)

TABLE VII

Table to indicate final diagnosis and treatment.

Diagnosis	No of Cases	Management.
Hirschprung's Disease	18	Duhamel's Optn 11 cases.
Idiopathi Megacolon	5	Myomectomy 1 drugs 3
Hypoaganglionosis	1	Myomectomy 1
Unconfirmed	11	(3 still have colostomies)

TABLE VII

Table of Complications experienced.

Complications	No of cases.
Strictures in anastomosis area.	4
Prolapse of colostomy	5
Poor anastomosis	2
Intestinal obstruction due to adhesions.	3
Incisional herniae.	1
Burst abdomen.	1
Perforation of bowel wall at biopsy.	1

INVESTIGATIONS and RESULTS

Table VI

INVESTIGATIONS	No of CASES	RESULTS
Plain Abdominal X-rays	13	No tentative X-ray diagnosis
Barium enema	22	15 (69%) Suggestive of Hirschsprungs.
* Both	7	——
Rectal and Colon biopsies	25	18 confirmed Hirschsprung's disease
Laparatomy.	24	
(Autopsy)	1	——

* 'Both' means Ba enema and plain abdominal X-rays were done on the patient.

Results.

Resections at definitive surgical treatment indicate that the majority of the cases had what is called 'short' segment aganglionosis. Only five had the resected Segments of colon measured 20cm. 17cm; 13cm; 16cm and 5cm.

Associated congenital anomalies in all the cases included a single case of malrotation of the whole gut. Many had presented with diverse conditions normally assosiated with Hirschprung's disease in a secondary role - recurrent chest infections (2) severe diarrhoea (2) Septicaemia (2) A case of megacolon presented with volvulus of sigmoid colon and volvulus of the small intestine with gangrene to which she succumbed.

Mortality

Out of 18 cases of proven Hirschprung's disease 3 died (Mortality of 16.7%) In all cases of megacolon, 5 patients died (Mortality of 14%)

No operative mortality was found in those done one form of definitive surgica treatment or another. 3 of the 5 who died had been done laparotomy.

Case I J.C. Age 2 weeks; Adimitted twice as acute abdomen. 2nd time laparo-

tomy and biopsy done also colostomy. Had Burst abdomen. Succumbed 3 days Postoperatively. Diagnosis Hirschprung's disease proved histologically.

Case II BM. Age 10 months. Child with poor health. Done rectal biopsy. Developed severe diarrhoea and died Diagnosis proved Hirschprung's disease.

Case III K.M. Age 5 days.

Had septicaemia at admission and acute abdomen. History suggestive of Hirschprung's disease Laparotomy, colostomy and biopsy done. Operative findings consistent with Hirschprung's disease and proven histologically. Died 2 days post-operatively.

Case IV IM. 12years Old.

Constipated since childhood with bouts of diarrhoea provisional diagnosis of megacolon in toxic state was made. In the ward developed acute abdomen, at laparotomy hypertrophic colon with volvulus of signoid colon and ileum and gangrenous segments. Biopsy by forceps was not conclusive at histology. Resected bowel showed normal ganglion cells. Died same day.

Case V (J.S.) 15 years Old.

Admitted with history consistent with intestinal obstruction. Had long standing history of problems with emptying of bowels since birth. Provisional diagnoses of megacolon made. Developed diarrhoea in the ward and died a day later.

At autopsy stercoral ulcers in grossly dilated and hypertrophic colon with perforation of bowel wall and peritonitis found.

At histology tissues were too autolysed to be of any diagnostic value.

DISCUSSION:

Hirschprung's disease is among those diseases which were thought to be rare in Africans. (Shepherd 1966. Katz 1966), but it has never been considered entirely absent. It is one of those diseases whose mortality in neonatal life and childhood is so marked that it may be thought to be rare. Indeed the incidence of a disease can only be determined by proper recording of diagnoses in hospitals and those seen at autopsy. It follows, then, that the less autopsy diagnosis are made the less the incidence of the disease. This is the state of affairs in our country. It would be fair at this moment to suppose that Hirschprung's disease's incidence is similar to anywhere else in the world. This fact can only be concluded by adequate statistics and at the moment, the above supposition works as a baseline for planing on diagnosis, investigations and management. The analysis on the 35 cases who were seen over a period of 5 years at The Kenyatta National Hospital is meant to compare the findings here and elsewhere where planning for this disease has been done and perfected.

Hirschprung's disease is compatible with adult life, but its mortality in childhood is very high (Hirschprung 1886) without surgical operation. The high mortality has been emphasized by: Zuelzer et al (1948), Burnard (1950); and Bodian et al (1951). This should be kept in mind.

Intestinal aganglionoses has been said to have anincidence of I in 20,000 -30,000 live births (Bodian et al 1951). Burnard (1950) estimated that for every 7000 live births 6 cases had aganglionoses. Other estimates are even higher 1 in 5,000 (Bodian et al 1963) Our situation is such that there is no reason to suppose any different figures from those when statistics are better than ours. Table X In the 18 cases of proven intestinal aganglionosis male predominance was evident M:F = 8:I. (16 males against 2 females) This male predominance has also been found in other series large and small McDonald et al (1954) (M:F 15:1) Bodian1951 (M:F =37:3) Whitehouse et al(1948) M:F = 10:1. The accepted average Sex ratios is M:F of 5:I.

No human race is free from intestinal aganglionosis Indeed, aganglionosis is not confined to the human race alone. A similar defect has been found in mice (a strain Piebald lAthal mutation). Lane(1966). These stains of mice have pigment deficiency and have been investigated extensively an attempt being made to draw any parallel between mice and humans in relation to aganglionosis. The importance of establishing the hereditary deficiency of ganglion cells may be important in marital counselling. The pattern of herdity has not been established in humans as in piebald mice; but the familial tendency of the disease is a recognised fact. It was noted early by Dalla Valle (1920). The same familial tendency was documented by HCDonald et al (1954). where they had a patient in their series who was the 6th victim of this malady in a family. Zuelzer & Wilson (1948) had a family of 15 with the disease. Bodian et al (1951) estimated that if the disease is in one child in a family a male child in the family is 1 in 5. It is the 'long' Segment which goes in families*. In this series no history was available to support any familial involvement. As far as tribal breakdown was concerned, the fact that there were more Kikuyu presenting with megacolon and Hirschprung's disease may be due to the nearness of this tribe to the Kenyatta National Hospital and the same may be said of the Kamba. Since the Hospital is a referral Hospital for the whole of Kenya, one can argue that probably facilities exist in the peripheral Hospitals so that very few of the many other tribe were available for analysis. If this is not true, then, very big number of cases of cases of Hirschprung's disease are not diagnosed. There is no reason to suppose that Hirschprung's disease is confined to 5 tribes in Kenya. More work is needed to create an awareness of the existence of the disease among the many cases of diarrhoea seen in children everyday in the

(* MARTIN 1972)

hospitals in the country. Nixon (1964) has attempted to define clinically the modes of presentation in the various clinical groups of Hirschprung's disease. The cases of Hirschprung's disease could be detected early and better if these groupings are comprehended and applied in various clinics.

Another aspect to be looked into in tribal incidence is the role the traditional medicine plays in Hirschprung's disease and other megacolon cases. In South Africa, the Bantus practise administration of enemata to children with varying abdominal ailments. (Van der Horst 1966). In Kenya. Such practise is not extensive among the tribes though purgatives (herbal) are widely administered. Whether there exists any relationship between the apparent low incidance of Hirschprungs disease and traditional practises in some tribes in Kenya, this will require more investigations.

The Kenyatta National Hospital serves mainly the African population, and therefore it would not show any racial representation in the disease, but there is no reason to suppose that Hirschprung's disease does not affect the other races in Kenya; - (or in Nairobi Specifically.)

Table IV,
represents the list of symptoms which were commonly found among the cases. In the last column figures from a series of proven cases of Hirschprung's disease by Daintree (1957) have been inserted for comparison: Abdominal distension was the most prominent symptom (77%) with constipation taking also a significant prominence (71%) In the proven cases these were (63%) and (69%) (Compare figures from Daintree where the two are 92% and 84% respectively). Vomiting was found in 40% of all the cases and 35% of Hirschprung's disease (Daintree's figure 76%) When these three symptoms are combined 23% of all cases had all while only 17% of Hirschprung's disease had all. (Daintree 58%). Diarrhoea is a common finding in cases of Hirschprung's disease. It takes two forms- mild form which follows a bout of days of constipation and the other form is usually severe (In some cases a terminal event) diarrhoea which is usually associated with ulceration of mucosa in the colon. (Note two of the patients who died had diarrhoea before they died). Diarrhoea in neonatal cases of Hirschprung's diseases takes a high toll of these cases. McDonald et al (1954).

Vomiting in 6 of the cases led to the diagnoses of acute intestinal obstruction and emergency surgery was indicated, otherwise it is an accompaniment of prolonged and severe constipation which is relieved by bowel emptying in most cases of Hirschprung's disease. Failure to thrive (5 cases) was found in 2 cases of proved Hirschprung's disease. This is a common finding in the population here and the causes are sundry. It is not a very marked finding in cases of Idiopathic megacolon. Incontinence of stool was among the group of 'others'. Two patients

were later proved to be Idiopathic megacolon and one hypoaganglionosis. Other symptoms were recurrent chest infections (Coughs). It is important to emphasize the significance of symptoms in Hirschprung's disease in the diagnosis. Typical cases present with constipation of varying severity and intermittence. Bill (1957) says Hirschprung's disease cannot be diagnosed in absence of constipation. Abdominal distension is essentially and logically a later development. In fact gross abdominal distension may not be there for example in cases involving the ileum and colon. It is better to discuss the various conditions which in one way or another resemble Hirschprung's disease at this juncture. Nixon (1966) has attempted to throw some light and establish some order in the complex field of these diseases. Briefly, he has on one end Hirschprung's disease and Chagga's disease representing those diseases which on histopathologic examination, show absent ganglion cells of the intestinal wall plexuses, without differentiating extensively the aetiology. It is known that Hirschprung's disease is congenital aganglionosis while chagga's disease is an acquired aganglionosis (Chagges 1909) due to infection with protozoan Trypanosoma cruzi. At the other end he put those conditions which, because of clinical feature resemble Hirschprung's disease, but on histopathology have 'normal' looking ganglion cells in the intestinal wall. These conditions have been described by many authors and surgeons with many names including PseudoHirschprung's disease, a word coined by Bevitch (1958). In this group may be included among others, the Idiopathic megacolon; the megasigmoid syndrome of the psychotic described by Krapf (1966) which has presentation and complications similar to Hirschprung's disease; the chronic constipation cases as described by Cookir and Cairdner (1960) and later by Nixon (1966) the segmental dilatation of the colon of Swenson and Rathauser (1959) (Hypertrophy also found) lastly, but not to exhaust the whole list, is that condition of complete adynamic bowel in neonatal cases, (Nixon 1966). In between these extreme cases a group of cases which show all Kinds of abnormalities (Morphological) in the ganglion cells exist. Among these are those premature infants whose clinical picture and indeed the radiological features on Barouma are similar to Hirschprung's disease. They become normal with time Warburtte has described them well. (Warburtte 1973 and Spencer 1966). The physiological explanation of these cases and their morphological behaviour has been well presented by Okamoto and Ueda in their studies on development of ganglion cells of intestinal wall in human foetuses (Ueda & Okamoto 1967). They showed the development and maturation of these cells goes on even after birth.

The other condition which falls into the abnormal ganglion cells is the so called hypoaganglionosis. Here it is the quantity of cells which has been said to be abnormal. Bentley (1966) has described these ganglion cells as those which are

seen in Hirschprung's disease at the so-called 'transitional' area - they are fewer in number. The Surgeons' interest in these cases is because they at one stage or another have been relieved by operations and because they present as causes of volvulus of the colon.
Their modes of presentation may differ with Hirschprung's disease in the time they appear in patients e.g. Idiopathic megacolon may or may not be associated with training a child to stool and appears at that age when this is done (Ravitch 1958). The history and symptons are therefore very important to the diagnosis.

It is my impression that symptoms as narrated by patients or relatives take a very important step in the process of diagnosing all cases of megacolon and Hirschprungs disease. An impression made at the history taking leads one to do the right investigations and may infact help in referring to the right specialist units in the hospital and if it is in the peripheral hospital to the referral centres.

The Age distribution of all patients are shown in the two tables III a and b. The youngest patient was 5 days old and was diagnosed Hirschprung's disease; while the oldest of the patients was 15 years Old. The latter died due to complications, but the diagnosis was finally uncertain at histopathology although she died of "typical" complication of Hirschprung's disease. The colon and sigmoid colon were grossly hypertrophic and dilated. The oldest with proven Hirschprungs disease was 8 years. The table III (a) indicates how many were seen yearly up to 2nd year of life In all cases over half were seen in the first year. It is worth noting that over 60% of Hirschprung's disease cases were seen after the first yearwhile only 40% were seen in their first year of life.
Table III(b) is meant to show a breakdown on how, after birth, we quickly saw the cases- so a weekly table of cases. Only 4 cases were seen in the first month (first 4 weeks) and only 7 cases in the first year. All other cases (11) were seen after the first 52 weeks. These are the proven cases of Hirschprung's disease. All cases show an increase with ages of the patients.

These tables would make logical sequence if table V is also taken into the discussion together with them. It tends to show, (its ambiguity apart) the relationship between the onset of symptoms as narrated by the relatives or patients and the age of the patients. 63% of all cases and 61% of proven Hirschprung's disease had symptoms "since birth" At the other end 22% of Hirschprung's diseas cases had symptoms setting in after one year. The obvious deduction here is that inspite of onset of the symptoms of Hirschprung's disease at a very early period in life the patients present at an advanced stage of childhood. Very few cases were seen in the neonatal period.

Nixon (1971) in his series of 155 cases of aganglionosis 70% are in the neonatal life and that these make the majority of the admissions in his hospital for intestinal obstruction. It is to be expected that a similar percentage should be seen in our hospital too, and if not, as the figures show, then were are missing the majority of the cases of Hirschprung's disease. The fact that neonatal admission forms the bulk of cases of aganglionosis in Europe and America rather than childhood admissions for the similar disease, makes the pattern of the disease as seen here different from the pattern there. The cause for failure to see the cases in the early life may be due to the populations ignorance about the disease or the medical personnel's unawareness about the disease or both. I think aganglionosis of the intestinal is a less well understood disease in the hospitals and among the populace. Efforts to educate all concerned is required.

The suspects are those who do not pass meconium or those who pass it inadequately (delayed). Some diseases are associated with aganglionosis eg. Down's Syndrome. Cases of unresponsive diarrhaeal diseases which normally present with abdominal distension and labelled electrolyte imbalance should be fully investigated (Nixon 1971). The paediatricians and Clinical officers who see most of these cases should take note of these facts.
It is only by early detection before these patients leave the maternity nurseries and afterwards in the Child welfare centres that we can reduce the mortality which has been put between 30% to 90% for aganglionosis (Zuelzer 1948; Barnard 1950; Fadian 1951). Investigations of Hirschprung's disease tend to be simplified with the better understanding of the disease. The contributions by Ehreinpreis (1946) on radiological role of diagnosing congental aganglionoses or any of the other allied conditions continues to play significant role up to now. The limitations and merits are now better studied. Modifications have been found to the basic methods - Swenson (1960) has emphasized the reliability of radiology. He said a radiologist is able to suspect aganglionosis in 15% of plain abdominal xrays, and, in his series, by Ba enema, 84% of cases were diagnosed; while in 16% of cases xrays were unhelpful. In the cases analysed here of the plain abdominal Xrays no tentative diagnosis was made while in the 22 Ba enemata done in 69% of the cases the radiologist was able to say they were suggestive of aganglionosis.

The limitations of Ba enema in diagnosis especially in prematures and neonates has been shown by Vanhautte (1973). The simplicity of the methods as compared to surgical methods especially in the very sick ones, is worth noting, but the dangers of water intoxication and death has been one of the cautionery measures to be undertaken. The enemas give information that cannot be given by biopsy reports. - The extent of the aganglionic segment and also the extent of the dilated segment. Both informations are important in the subsequent surgical management of the cases depending on the methods adopted

in the management (Vide infra).

My impression is that more use of radiology as a means of diagnosing cases of congenital aganglionosis or other megacolon diseases should be made. It should be a logical use of these methods in all suspicious cases. The other method of diaginosis that was used was rectal biopsy. 25 patients were done rectal biopsy in the conventional method (Swenson et al 1955; Nixon 1971) except 3. In these 3 Rectal biopsy forceps were used. The results of the three specimens were inadequate for the pathologists to give conclusive diagnosis. This method and punch biopsy are not popular. Full thickness bowel wall specimen has been the popular method of dignosis. By biopsy only can the aganglionosis be demonstrated, for diffeentiation of cases into the 3 categories - aganglionosis, hypoaganglionosis and 'normal' ganglion cells - it must be done and done properly. Biopsy could also be done at laparotomy. The aims are for diagnostic purposes and for establishing the length of aganglionosis in the sigmoid colon or higher by 'markers' for future treatment by resection (24 cases in the series were done laparotomy and biopsies taken, but in only 3 was the procedure primary. In the others the rectal biopsy had been done and laparotomy was not fordiagnostic purposes only).

Rectal biopsy is the only reliable method of diagnosis of aganglionosis and especially in neonatal period where radiology is unreliable (Swenson 1960).

One patient whose clinical diagnosis and autopsy findings were consistent with Hirschprung's disease was not at histology conclusively diagnosed as such. Autopsy is an unfortunate method of diagnosis. The discussion on methods of diagnosing Hirschprung's disease and other allied disorders could not be complete without mentioning other methdds though they are not at our disposable at the moment.

Rectal pressure studies have revealed special features of cases of aganglionosis. The method has been popularized in some centres after the initial studies by Lawson and Nixon (1967). The easiness of the method might probably make it popular, but more studies will be required in our centres here.

Rectal biopsies by suction method is another more popular method in Europe. The specimen consists of mucosa and submucosa and excludes the muscular layers and usually histochemical stains are used besides the ordinary staining methods. The method requires pathologists conversant with the interpretation requiring only these bowel wall layers (Noblett and Campbell 1969).

It is agreed that the method is less traumatic to the patients. However, it requires special instruments and experience in its use.

The methods of investigation and diagnosis can't be completely and wholly discussed without paying particular attention to the importance of physical Examination and history in the diagnosis of these diseases. The use of these methods is even more significant in our country where not all hospitals are exposed to the special facilities, and even where they are, the qualified personnel to interprete them are scarce or absent.

The investigations in all 35 cases led to the confirmation of aganglionosis in 18 cases; idiopathic megacolon in 5; hypoganglionosis in one which left out 11 cases unconfirmed (5 were lost to follow up in surgical Clinics after they were referred by Paediatricians; 6 patients were dispersed as follows:- One died before biopsy (NO.P.M. was done) 3 aborted for colostomy and biopsies but the latter were inadequate for diagnosis; One was done anal dilalation and the other is scheduled for operation at the age of one year.)

In the follow up clinics, attendances are mainly by those who still have colostomies (4 of aganglionosis and three with unconfirmed diagnosis so far) and most of the other appear in the Wards with various complications. In general, follow up is inadequate to assess the effectiveness of operative management in Hirschprung's disease as well as drug therapy in the cases of idiopathic megacolon which have been so managed. The overall analysis of a all cases who were diagnosed, by one method or another, indicates certain failures in the system either the referral of cases or even in management. Many experienced workers in this field say no patient suspected of aganglionosis should go home without confirmation of diagnosis or even treatment (Nixon 1971). This is a logical stance to take in all cases of aganglionosis or other allied diseases. The treatment and subsequent management is determined by the final diagnosis. The seriousness of the disease (aganglionosis) without surgical treatment must be understood. The disappearance of those cases who are sent for follow up in outpatient clinic from subsequent follow up is common in this hospital and country. Therefore when the patient is first seen he should be fully investigated and management planned according to the principles of the surgeon in charge. The other differential diagnosis entertained in some cases were meconium ileus; faecal impaction, toxic megacolon volvulus and malrotation of the gut. 9 patients were diagnosed at laparotomies.

The table VII, besides, showing the final diagnosis shows also how the cases were managed. In the 18 proven cases 11 of them were done definitive operation - all of them Duhamel's operation. In all the cases 60% were done preliminary colostomies . Two cases were done myomectomies (One hypoganglionic

case and one idiopathic megacolon). 3 cases of idiopathic megacolon were managed on medicines only-

Colostomies are indicated in most cases of congenital aganglionosis as a temporary measure except in those cases who are managed by primary definitive operations. Colostomy is also indicated in those cases of idiopathic megacolon where signs of intestinal obstruction are found. In these cases t the colostomy is life saving and it may be a temporary measure and in some cases a necessity for life (One of the cases was in latter category) with such obvious indications, then, the problem of the attending surgeon is not the indications, but the appropriate site for the colostomy in a patient. It is well to caution that colostomy operation is not a small operation considering the type of cases the surgeon is dealing with. A poorly placed colostomy may make definitive surgery technically difficult. A treacherous colostomy is that pl aced in the 'transitional' segment where ganglion cells are scanty and bowel motility poor - the colostomy does not work adequately.

In 18 cases of those who had colostomies they were sited at the right side of the transverse colon. This site has been claimed to be ideal, but it has also been argued that in neonates siting the colostomy there renders the distal end less well developed especially the vascular components due to disuse atrophy. At the time of definitive treatment there may be difficulties in bringing down the colon for anastomosis. This is logical, but practically the difficulties have been overcome. In 3 cases the colostomies were located in the sigmoid colon. The method has the advantage over the above in that the whole functioning colon is given opportunity to fully develop, but has the disadvantage that it has to be dismantled at the time of the definitive operation which is not normally done in transverse colostomy. In cases of total colonic aganglionosis ileostomy is indicated.

Nixon (1971) has discussed the method of doing colostomies in children. The colostomy should be stitched mucosa to skin and not to use loop-and-rod method due to its subsequent complications in children. In all cases of colostomies, regular anal dilatation is mandatory to keep the anus patent for use in subsequent anastomosis.

The definitive treatment for Hirschprung's disease is surgical operation. Many forms of definitive treatment exist and they all have their own proponents and opponents. The choice belongs to the surgeon depending on his experience. In all 11 cases Duhamels operation was done-

(Duhamel 1960). Many other which one could choose from are such as the original one - Swenson's pull-through operation (Swenson 1948; Swenson 1950); Soaves endorectal submucous pull-through (Soave 1964) Martius operation (Martin 1962)

States operation (state 1952) All these have a common denominator that the the narrowed segment is resected or excluded and continuity is established by anastomosis. The different ways in which the anastomosis is effected is what contributes to the differences in these methods. Among these the Duhamels' operation has been claimed to be less involving than the others. Technical difficulties apart all these methods are essentially the same. It is the experience gained when one surgeon uses one method that determines the choice.

The method which now has been very much favored in selected cases is the myomectomy or (2 cases had this). Sphincterectomy. (B.Duhamel 1966; Bentle 1966) In the two cases this was met with success - one case of hypoaganglionosis. In some cases of idiopathic megacolon and in those cases of Hirschprung' disease who show, on Barium enema, the termed " ultrashort segment" aganglionosis he also manages those with aganglionosis of distal third of the rectum by what he called P.E.A.R.M. (posterior Excisional Ano Rectal myomectomy). In this, only the internal sphincter is excised - a strip of 5 mm width and 10 cm length extending upwards to the rectum. It is said that by so doing he has not only cured these cases (follow up of 9 years) but made a diagnosis of 'ultrashort' segment thus.

The method in selected cases has been found successful and requires trial here in those selected cases as it is within the capabilities of the inexperienced surgeon (in the other methods) and it is curative. Two importa aspects of management should be mentioned. In these cases who are found fit and have aganglionosis to do a colostomy as a preliminary measure awaiting definitive later is the method adopted by many surgeons, but it has been argued that in fit children one could proceed with definitive surgery after necessary colonic preparations without colostomy first. Swenson (1960). advocates colostomy and later definitive operation. For neonates with enterocolitis, which is the killer in Hirschprung's disease, surgery, is co contraindicated. According to his experience medical treatment should be undertaken. The mortality in neonatal period when one stage definitive surgery is performed is prohibitively high.

In this series of cases, according to the resected segments of the colon the 'short' and 'moderate' segments were the commonest. (See results). Else where cases of total colonic aganglionosis have been seen by the doctors but with improvement on selection and diagnosis of our cases in neonatal period the problems of long or total aganglionosis of colon and ileum will be seen hereto. An ingenious operation has been devised by Martin (Martin 1972). In the operation the aganglionic colon is utilized for its absorptive purposes

and is not resected. A long side-to-side anastomosis is made between the aganglionic and the ganglionic ileum, the latter being useful in effecting motility and propulsion of contents. The method has worked sucessfully in an acceptably large series.

Hirschprung's disease has a high mortality when medical care is scarce. The mortality as stated earlier on is high in neonatal period where figures vary from author to author and country to country. Nixon (1964) says the mortality is about 50% or higher at the neonatal period. State (1952) said before the antibiotics the mortality ranged from 37% to 79% higher figures are even quoted for this disease. The cases who were seen in the hospital, 3 proven cases of anganglionosis, and 2 with inconclusive diagnosis died. In operated cases (definitive) there was not mortality and the morbidity in form of complications experienced in couse of total management (in diagnosis colostomy and definitive treatment) are relatively low. and are no more than those met in cases of laparotomy for other conditions. Inspite of poor follow up the success of the management of these cases was obvious at the time of discharge from the hospital. The causes of mortality in cases of Hirschprungs disease is severe enterocolitis. These cases are managed as gastroenteritis but the condition is recurrent, and this is where the diagnosis requires suspicion rather than looking for typical symptoms. Those who have long segment aganglionosis which at times includes the ileum have even higher mortality at best of the hands - greater than 65% (Martin 1972).

The other cause of mortality which is iatrogenic in nature is at enemas to evacuate the bowel. Before the hazard of using soap water or even 'ordinary water for rectal enema was realised cases of sudden deaths were many. The explanation for this is that water is introduced to the colon which is dilated and so increased surface area for absorption. When the child strained even more water is absorbed. They are said to die of water intoxication or "Hypronatraemic shock" A simple water enemas is contrandicated in aganglionosis (Lilie 1969). The same principle expains death after barium enema where much of the substance is instilled into the dilated rectum. For enema 'normal' saline is recommended.

Due to low mortality in definitive surgery in cases of aganglionosis it has been said that no child should be discharged out of the hospital without definitive treatment or a colostomy. (Nixon 1971). Swenson (1960) says in neonatal period the children should be sent home on colostomy until they are 9-14 Kg. in weight. He also argues that in those who present with symptoms of intestinal obstruction, if the diagnosis can be established as aganglionosis, they do not requre emergency surgery. 15% of his 200 cases required emergency

surgery. (In this series 23% required emergency surgery) On the question of age at which the definitive operation should be done (without considering the weight) State (1952) has cautioned the somatopsyhic consideration when dealing with these children. He emphasizes that these children present with personality and behavioural problems. This fact, then calls for dealing with aganglionosis as early as the child is physically fit to under go the operation.

Hirschprung's disease must be diagnosed or suspected as early as the children are in a maternity nursery. The management must start then. What ever method the surgeon adopts no child should be sent home without a diagnosis, a temporary measure or definitive operation having been done. It is by so doing that the mortality attached to this disease without proper management could be reduced. As Nixon (1964) has said it is only those with 'mild' aganglionosis who make it to childhood.

S U M M A R Y.

An analysis of 35 patients seen at The Kenyatta National Hospital over a period of 5 years has been done. 18 cases have been shown to have intestinal aganglionosis of 'short' to moderate' segments. The incidences in Sex and tribe have been analysed and sex incidence found to be equal to other series. It has been coucluded that inspite of only having 5 tribes in the series there is no reason to assume that other tribes not represented do not suffer from aganglionosis. The prevalent presenting symptoms have been shown to be similar to those experienced by authors in other series and these are constipation, abdominal distension vomiting. Diarrhoea has not been a 'remarkable symptom although its importance elsewhere has been emphasized. In the investigations, the available and used methods have been shown to contribute to the diagnosis, the important ones being Ba-enema and rectal biopsy. In the age incidence the disparity in time between onset of symptoms and the time the patients seek help has been high-lighted and has demonstrated that this forms significant difference in the patten of the disease as seen in America and Europe and as it is seen here. Emphasis has been put on early diagnosis to check on martality associated with the untreated cases.

The analyses has also established that although there are many diseases, allied to intestinal aganglionosis "idiopathic' megacolon is important here where 5 of the 35 cases were consistent with clinical and histological diagnosis. The remaining unproven cases, though symptomatically and clinically shown to be aganglionosis or other allied disorders, were unfortunate failure and that this should be avoided by all means. In the analysis of methods of management it has been established that the mortality is low or absent except in the poor risk cases. (3 patients out of 18) and (in all 5 out of 35)

The importance of colostomy in all cases has been shown to be popular while myomectomy has been done successfully in 2 cases.

The importance of clinical examination and history of these disease in establishing the diagnosis has been borne out and emphasized and especially in our circumstances in peripheral hospitals where specialised facilities are absent.

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