PATTERN OF AGANGLIONIC MEGACOLON IN CALABAR, NIGERIA

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Hirschsprung's disease, a congenital condition resulting in functional intestinal obstruction, was seen in 21 children at the University of Calabar Teaching Hospital (UCTH) between January 1996 and December 1998. Late presentation of patients to hospital was observed and was attributed to ignorance and poverty. Patients presented only when repeated herbal enemas no longer offered relief. The main complaints were abdominal distension, constipation, wasting, diarrhoea and growth retardation. Surgical resection of the aganglionic segment and anastomosis is the treatment of choice and this procedure decreases mortality and morbidity. Palliative transverse colostomy was considered essential as it protects the anastomosis. Early diagnosis of this disease in our environment can only be achieved by means of a public awareness campaign. This will prevent many of the fatal complications associated with this condition.

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Hirschsprung's disease is the main genetic cause of functional intestinal obstruction, with an incidence of 1/5 000 live births.[1] It is characterised by a sustained contraction of the large intestine which may involve a long or short segment.[2] The commonest site of involvement is the rectum up to the distal sigmoid, but 20% are shorter than this and some longer, with 4% of cases spanning the entire colon and extending to the small intestine.[3] It is a developmental disorder, a neurocristopathology, and is characterised by the absence of the ganglia along a variable length of the intestine. Surgical resection of the aganglionic segment with anastomosis is the treatment of choice[4] and surgical approaches have dramatically reduced mortality and morbidity. Early diagnosis is vital[3,4] so as to prevent necrotising enterocolitis which is a known fatal complication,[5] especially in the neonatal period.

To the best of the author's knowledge, the pattern and presentation of Hirschsprung's disease in children in south-eastern Nigeria has not yet been documented. The aim of this survey, therefore, was to evaluate the age and sex incidence, clinical presentation and other features of the disease in south-eastern Nigerian children admitted to the University of Calabar Teaching Hospital (UCTH), Nigeria.

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MATERIALS AND METHODS

This is a prospective study of children admitted and treated for Hirschsprung's disease during the period January 1996 through December 1998. The UCTH is the only tertiary health institution serving the whole of the south-eastern part of Nigeria, with a population of about 7 million (1991 Nigerian census).

All children under the age of 15 years who were histologically diagnosed and treated for Hirschsprung's disease at the UCTH during the 3-year period were included in the study. Relevant information obtained from the parents included age, sex, socio-economic status, symptomatology, time of onset of illness and duration of illness, remedies applied before presentation at UCTH and complications, if any.

Relevant laboratory investigations included haemogram, stool microscopy, barium enema and rectal biopsy for histopathological examination.

Treatment in all cases, irrespective of the type, started with fluid replacement therapy, colonic irrigation using normal saline, and in some cases, palliative colostomy. In cases presenting with enterocolitis, antibiotic treatment was instituted. Definitive surgical correction was effected after stabilisation of the general condition of the patient.

RESULTS

During the period of this study, 21 children were confirmed as having Hirschsprung's disease and treated at the UCTH. There were 17 boys and 4 girls, giving a male/female ratio of approximately 4:1. Only 1 patient presented under 12 months of age, while a majority (14 cases, 67%) presented between the ages of 1 and 6 years (Table I).

On the basis of clinical presentation and rectal examination, two types of symptom complex in Hirschsprung's disease were identified.

Type I. The patient is wasted with insidious abdominal distension and visible colonic peristalsis, but is alert and well hydrated and life is not threatened. This is the chronic form of presentation.

Type II. The patient presents with rapidly progressive abdominal distension, spurious diarrhoea, necrotising enterocolitis and rapidly deteriorating cardiovascular status, with eventual cardiovascular collapse. This is the acute form of presentation.

Table I. Age and sex distribution in 21 children with Hirschsprung's disease at UCTH

	Age (years)						
	Under 1	1 - 3	4 - 6	7 - 9	10 - 12	13 - 15	Total
Male	1	6	5	3	2	0	17
Female	0	2	1	1	0	0	4
Total	1	8	6	4	2	0	21

The major complaint in the UCTH patients was constipation with progressive abdominal distension. In 6 patients medical assistance was sought because repeated herbal enemas could no longer relieve the constipation and progressive abdominal distension. Repeated herbal enemas had earlier helped in cleansing of the colon, hence preventing enterocolitis. Nine patients presented with spurious diarrhoea and abdominal distension (Table II). A majority of the patients (15, 72%) presented with varying degrees of wasting and growth retardation, such as stunted growth and poor weight gain.

Fifteen of the 21 children were of low socio-economic background, their parents being subsistence farmers and artisans.

The age of onset of symptoms varied. Patients with the type I pattern of the disease reported to hospital later than patients with the type II pattern, as the type II pattern was acute demanding urgent attention. In 6 patients (28%) constipation started within the first year of life, with 3 patients (15%) constipated from the neonatal period. In 9 cases (40%) constipation had occurred within the first 3 years of life.

Table II. Symptomatology of Hirschsprung's disease in 21 children at UCTH

Signs/symptoms	No. of cases (%)*
Abdominal distension	21 (100)
Constipation	12 (57.1)
Wasting	11 (52.4)
Visible colonic peristalsis	10 (47.6)
Spurious diarrhoea	9 (42.9)
Growth retardation	9 (42.9)
Palpable colonic faecal stones	5 (23.8)

* Some patients had multiple symptomatology.

The aganglionosis seen in this survey involved the rectosigmoid segment in all cases. In 2 patients the aganglionosis affected an ultra-short segment of the rectum.

As a result of clinical evaluation and investigation, 13 patients with the type I pattern had transverse loop colostomies and were referred for definitive surgery. Patients with type II pattern of the disease (8 cases) were treated with colonic saline irrigation for deflation of the colon before transverse loop colostomy was instituted as palliative treatment.

Two patients with type II disease developed fulminating enterocolitis with fatal consequences.

Twenty-one patients were operated on: 11 patients using the Swenson approach, 5 patients using the Duhamel method, and 3 patients using the Soave method. In 2 patients with an ultra-short segment of the disease Lynn's dorsal myectomy was done because of the ease of application (Table III).

In the follow-up period of 6 - 18 months, 10 patients had satisfactory bowel habits and were continent. Three patients complained of constipation, which was initially mild but insidiously worsened to necessitate the use of enemas. Three patients had total incontinence and 2 patients were lost to follow-up.

Table III. Surgical procedures and associated complications seen in Hirschsprung's disease in 21 children at UTCH

Surgical procedure	Patients (*N*)	Type of disease		Complications				
		I	II	Anastomotic leak	Wound infection	Constipation	Faecal incontinence	Enterocolitis
Swenson	11	7	4	1	2	2	2	2
Duhamel	5	3	2	1	1	1	-	-
Soave	3	3	-	-	1	-	1	-
Myectomy	2	1	1	-	-	-	-	-
Total	21	14	7	2	4	3	3	2

DISCUSSION

For a long time Hirschsprung's disease was thought to be very rare, but with increasing awareness and ease of diagnosis worldwide the recorded prevalence rate has increased.[5] Nevertheless, in a society like ours where over 60% of the populace cannot afford hospital treatment and have no access to tertiary-level health care, some cases of Hirschsprung's disease are still likely to go undiagnosed. It may therefore mean that Hirschsprung's disease can be an important contributory factor in childhood mortality and morbidity in our environment.

Most of the children in our series were reported after the infancy period — only when they had developed irreversible intestinal obstruction, abdominal distension or enterocolitis did the parents seek hospitalisation. It appears that the widespread use of herbal enemas, which have cleansing properties, tide these infants over the neonatal period, but the traditional concoctions tend to become ineffective after the first 12 months of life. This may therefore explain the late presentation of patients for treatment.

The picture is at variance with the presentation in Europe[6] where the majority of patients present early, largely during the neonatal period.[3,5] Orthodox enemas, with their cleansing properties,[7,8] help to prevent fulminating enterocolitis. Perhaps it may be assumed that frequent traditional enema application among the populace also prevents enterocolitis and therefore afflicted children can survive the early months.

The sex distribution in this survey is in consonance with the pattern elsewhere.[6] There is a male preponderance of approximately four males to one female.

Patients were operated on after preliminary stabilisation of their fluid and nutritional status. The policy of our unit has been to institute a protective transverse colostomy in all cases. This procedure may be open to criticism but the experience of some investigators[3] has shown that this approach prevents anastomotic leaks, encourages rapid healing of the anastomosis and can be used to assess the functional status of the anastomotic site. Anaerobic septic complications of transverse colostomies were not noticed, apparently because of the policy of using broad-spectrum antibiotics intra- and postoperatively. A palliative transverse colostomy is essential, especially in our locality. Known associated complications arising from premature, outright resection and anastomosis such as anastomotic leaks, strictures, peritonitis, bowel necrosis and wound infection[3,4,9] can thus be avoided.

Late reporting of patients to hospital may be attributable to ignorance and poverty; 2 patients died in the series as a result of fulminating enterocolitis. Overall, 10 patients or 48% of cases had satisfactory bowel habits, although it may be argued that the follow-up period in this preliminary report of the survey was not long enough. Three patients had various degrees of obstruction and 3 patients had faecal incontinence. Results from other centres[6,10] have shown a similar trend but it was observed that with time these functions improve.

In conclusion, therefore, public health education is necessary with regard to seeking early medical attention and the exact incidence of Hirschsprung's disease.

This should enhance early surgical correction and prevent fatal complications.

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