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Hirschsprung's Disease at the Lagos University Teaching Hospital

SD ADEYEMI*, B OLAYIWOLA† AND JT da ROCHA-AFODU††

Summary

Adeyemi SD, Olayiwola B and da Rocha-Afodu JT. Hirschsprung's Disease at the Lagos University Teaching Hospital. *Nigerian Journal of Paediatrics* **1986; 13:7.** Sixty four children who presented with Hirchsprung's disease between 1975 and 1984 at the Lagos University Teaching Hospital, were studied. Twenty-seven (42.2%) of the 64 presented with intestinal obstruction requiring emergency right transverse colostomy in the neonatal period. The remaining 37 (57.8%) presented later with chronic constipation. Enterocolitis and peritonitis did not feature as forms of presentation or complications in the series. A steady rise in the proportion of newborns with Hirschsprung's disease was observed during the period. Diagnosis was established by barium enema and rectal biopsy in all the 64 cases. Only 30 of these children have had definitive surgery; 16 of them had modified Duhamel's procedure, 12 had Swenson's procedure and two had Soave's procedure. Follow-up periods ranged between 3 and 96 months, with a mean of 45.6 months. The results, classified into good, satisfactory and poor, were encouraging for Duhamel's and Swenson's procedures, but poor for Soave's procedure. The mortality in the series was 8%; three of the five deaths occurred following colostomy and the remaining two were associated with definitive surgery.

Introduction

ALTHOUGH discussion of Hirschsprung's disease in developed countries has shifted from considerations of clinical presentation and management to more sophisticated researches, the pattern and clinical peculiarities of this disease as it presents in the developing world including Nigeria, are yet to be fully described. In addition, the problems posed by our low state of socio-economic development to the proper management of this disease, make it desirable for individuals or groups who have gained appreciable experience in tackling these problems to share it with others in similar situation. This way, not only will the pattern of the disease in our environment emerge, it should be possible to determine what is achievable given our special and peculiar circumstances.

Lagos University Teaching Hospital, Lagos

Department of Surgery

* Consultant Paediatric Surgeon
† Chief Resident
†† Consultant Surgeon

Correspondence : Dr SD Adeyemi

As far as we are aware, all previous studies of Hirschsprung's disease in West Africa have involved small numbers of patients[1-6]. Solanke[1] in 1968, reviewed intestinal obstruction in Ibadan and listed only 7 cases of Hirschsprung's disease seen over a ten-year period. In another 10-year study of intestinal obstruction from the Lagos University Teaching Hospital, Olumide *et al*[2] identified 14 children with Hirschsprung's disease while another review from the same hospital by da Rocha-Afodu and Adeoba reported 15 cases of the same disease also seen over a 10-year period[6]. The experience in Zaria as reported by Momoh[3] consisted of 21 cases of suspected Hirschsprung's disease seen between 1972 and 1980; only 7 of these were investigated with barium enema and none was said to have had a rectal biopsy to confirm the diagnosis. Hirschsprung's disease did not feature prominently in the reviews of intestinal obstruction from Ghana and Sierra-Leone[4 5]. It is against the foregoing background that we wish to report our experience with 64 cases of Hirschsprung's disease seen at the Lagos University Teaching Hospital (LUTH), during the ten-year period, 1975–1984.

Materials and Methods

The records of children who were treated for Hirschsprung's disease at LUTH between 1975 and 1984 were analysed with respect to yearly distribution, age at presentation, sex, mode of presentation, investigations and management. The results of the definitive pull-through procedures were graded as good, satisfactory or poor, according to the classification suggested by Nielson and Madsen[7] as follows:

(*a*) "good", if the patient had no complaints, and passed normal stools spontaneously with a maximum interval of 2 days;

(*b*) "satisfactory", if the patient had only minor bowel disorders such as constipation which could be managed by laxatives but never required enemas, occasional diarrhoea, occasional slight abdominal distension and occasional soiling; and

(*c*) "poor", if there were frequent distensions of the abdomen and severe constipation requiring treatment with enemas.

Results

The 64 children studied consisted of 52 boys and 12 girls, a male to female ratio of 4:1. Yearly distribution of cases did not show any definite trend but the proportion of newborns rose steadily (Table I).

Clinical features

Twenty-seven (42.2%) of the 64 children presented in the neonatal period, 20 (31%) between the ages of 5 weeks and 12 months while 11 (18.9%) were aged 13 months to five years. The remaining 6 (9%) children presented between the ages of 6 and 15 years (Table II).

TABLE I

Yearly Distribution in 64 Cases of Hirschsprung's Disease

Year	*Total No of Cases*	*No of Newborns*	*% of Total*
1975	3	0	0
1976	7	1	14
1977	7	1	14
1978	9	2	22
1979	5	1	20
1980	8	3	38
1981	6	4	67
1982	11	8	73
1983	6	5	83
*1984	2	2	100

*Study period extended to April 1984

TABLE II

Age at Presentation in 64 Cases of Hirschsprung's Disease

Age	*No of Cases*	*% of Total*
0 - 4 weeks	27	42.2
5 weeks - 12 months	20	31.2
13 months - 5 years	11	17.2
6 - 10 years	3	4.7
11 - 15 years	3	4.7
Total	64	100.0

All the newborns presented with intestinal obstruction requiring urgent colostomy for decompression while the remaining children presented with chronic constipation. None of the cases presented with enterocolitis or bowel perforation.

Surgical procedures and complications

The complications of colostomies in the 27 newborns were wound infection (22 cases), prolapse (18 cases) and skin excoritation (16 cases), while retraction and gangrene occurred in one patient each. Three neonates died following colostomy.

Biospies taken during surgery revealed that a majority of the children had aganglionic segments extending to the recto-sigmoid. One child who later died of aspiration, had involvement of most of his large bowel.

Thirty (46.8%) of the 64 children had had definitive surgery in the form of an abdomino-perineal pull-through procedure at the time of this study; the remaining 34 children were awaiting definitive surgery following the initial construction of colostomy. Sixteen (53.3%) of the 30 who had surgery had modified Duhamel pull-through[8], 12 (40%) had Swenson pull-through[9] and 2 (6.7%) were treated with Soave endorectal pull-through[10].

Early and late complications associated with the different types of pull-through procedures are shown in Table III. The wound infection rate ranged from 33% (Swenson's) to 50% (Soave's); the rate was 38% in the group that was treated with Duhamel pull-through. Soiling was another frequently encountered late complication occurring in 53% of the children treated. It occurred in 41% of the Swenson's group, 56% of the modified Duhamel group and in the 2 children who had Soave's pull-through. There was one death in each of the modified Duhamel and the Soave groups. The overall mortality in the entire study including the three colostomy-related deaths was five (8%).

TABLE III

Complications of Definitive Procedures in Hirschsprung's Disease

Complications	*Type of Procedure*			*Total*
	Modified Duhamel (16)	*Swenson* (12)	*Soave* (2)	(30)
Early				
Wound infection	6	4	1	11
Chest infection	2	1	-	3
Burst abdomen	2	-	-	2
Anastomotic dehiscence	-	1	-	1
Urinary tract infection	-	1	-	1
Perineal abscess	-	1	-	1
Late				
Faecal impaction	2	-	-	2
Faecal incontinence	-	-	2	2
Soiling	9	5	2	16
Anastomotic stenosis	1	-	2	3
Anastomotic dehiscence	-	1	-	1
Incisional hernia	2	1	-	3
Adhesion/ Obstruction	-	1	-	1

Figures in parentheses = Number of patients on whom the procedure was performed.

Outcome

The duration of follow-up and results of surgery are summarized in Tables IV and V. Eight (26.6%) of the 30 children who had definitive surgery had been followed-up for 61 to 96 months. Another 8, including the two who had Soave procedure, were followed-up for 25–60 months, 5 (16.6%) for 13–24 months; another, 6 (20%) for 6–12 months and the remaining 3 (10%) children were seen for less than six months following discharge from hospital (Table IV).

Using the classification suggested by Nielson and Madsen,[7] the two children in the Soave group had poor results (Table V). In the Swenson group, 5 (41.6%) of 12 children had good, 6 (50%) had satisfactory and 1 (8%) was judged to have had a poor outcome. Five (31%) of the 16 children that had modified Duhamel pull-through had good results while 9 (56%) and 2 (12.5%) children had satisfactory and poor results respectively. Overall, 10 (33%) children had good, 15 (50%) had satisfactory, while 5 (17%) had poor results.

TABLE IV

Follow-up in 30 Cases of Hirschsprung's Disease who had pull-through Procedures

Procedure	*Duration of Follow-up (months)*				
	<6	6-12	13-24	25-60	61-96
Soave	-	-	-	2	-
Modified Duhamel	2	2	3	3	6
Swenson	1	4	2	3	2
Total	3	6	5	8	8

Average follow-up period was 45.6 months.

TABLE V

Results of Pull-through Procedures in 30 Cases of Hirschsprung's Disease

Procedure	*Good*	*Satisfactory*	*Poor*	*Total*
Soave	-	-	2(100)	2
Swenson	5(41.7)	6(50)	1(8.3)	12
Modified	5(31.2)	9(56.3)	2(12.5)	16
Total	10(33.3)	15(50)	5(16.7)	30

Figures in parentheses represent percentages

Discussion

When compared with previous reports, it is tempting to conclude from our findings that there has been a rise in the incidence of Hirschsprung's disease in Nigeria and West Africa. The 64 cases of the disease seen over a 10-year period in the present review represent a significant rise compared with 14 and 15 cases seen over similar durations of time in the past.[2,5] It is however, more likely that these observed increases are more apparent than real and due to increased paediatric surgical activity and an increased awareness on the part of parents and the primary physicians. A similar apparent increase in the incidence of Hirschsprung's disease was also evident in Ibadan where a 1981 review showed that there were 6 children with the disease during a 12-month period[11] compared with 7 cases spread over a period of 10 years two decades earlier[1].

The change in the pattern of presentation may also be due to the same reasons. The days in which Hirschsprung's disease was thought of as mainly a cause of constipation in an older child or even an adult as was the case in one of Solanke's[1] patients, are disappearing. It is

gradually becoming mainly a disease of the newborn in whom it presents as intestinal obstruction rather than constipation. This view is supported by the large percentage of newborns in this review and in Momoh's series.[3] Furthermore, perforation of the bowel with peritonitis as a mode of presentation or complication of the disease[6 12] was absent in this series. Although enterocolitis mimicking gastroenteritis did not feature in this review, there were some paediatric patients seen during this period with diarrhoea and abdominal distension[3 13] and in whom Hirschsprung's disease was suspected; such cases either died or disappeared before confirmation with a rectal biopsy could be carried out. Gastroenteritis is quite common in children in developing countries and it is possible that many cases of Hirschsprung's enterocolitis are sometimes, treated as cases of gastroenteritis, the error never being discovered if the child dies and no autopsy is performed.

Whenever possible, a child suspected to have Hirschsprung's disease should be investigated with barium enema and rectal biopsy before treatment is commenced. However, these investigations were not always possible on an emergency basis and were therefore, often deferred in our patients with obstruction until sometime after a life-saving right transverse colostomy had been performed or in some cases, following saline enema decompression. There are some pitfalls in using barium enema as a confirmatory investigation especially in neonates.[14] For example, in the left colon syndrome of babies of diabetic mothers, the barium enema appearance is similar to the picture in Hirschsprung's disease. In this study, rectal biopsy was positive in 12 neonates in whom barium enema was equivocal. Anorectal manometry[16] and measurement of acetyl cholineesterase activity in aganglionic specimen[16] have been used as additional diagnostic tools in some centres, but these were not available in our hospital.

The newborn with Hirschsprung's disease is usually managed with a colostomy until it weighs 8–10kg when definitive operation is carried out. Our children carry colostomy dressings rather than bags which are hardly available.[17] The older child who presents with Hirschsprung's disease also gets a colostomy which facilitates bowel preparation but in most cases, the child remains in hospital until a pull-through is performed shortly after. Abdominal perineal pull-through as the initial treatment of Hirschsprung's disease, particularly in the neonate, is not practised at this centre. The only complication of colostomy which was frequently a source of anxiety to parents in the present study was prolapse which occurred in two-thirds of the patients. When it occurred, the child was readmitted and efforts geared towards performing the definitive surgery early.

Follow-up, as is generally the case in Nigeria, was not satisfactory in the present series in which the follow-up period ranged from 3 months to 96 months with a mean of 45.6 months. The results of Swenson's and Duhamel procedures were encouraging. Soiling was a major late complication of both procedures in this study but it has been observed that the problem tended to lessen as the child grew older and with careful dietary selection, such as exclusion of beans and bean-containing food items from the diet. With regard to the two children that died after resection, the one with near total colonic aganglionosis would not have survived given our present level of medical care. Avoidable mortality in the resected group was therefore, 1 (3%) which is comparable to the one death in 34 resected cases reported by Cram.[18]

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