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Research Article

Management Of Hirschsprung's Disease Revisited: Single-Stage Versus Multi-Stage Operation: A 15 Year Retrospective Review Of Cases Seen At The University Of Nigeria Teaching Hospital Enugu, Nigeria.

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Abstract

Background: Hirschsprung's Disease (HD) is a cause of intestinal obstruction in children. In recent times operative management has transited from multiple to single stage procedure in developed countries. However, in resource-poor settings of the developing countries, this transition is slow to occur, with most of the patients still requiring multi-stage operation due to late presentation.
Objective: We retrospectively reviewed all cases of HD managed over a period of 15 years at the University of Nigeria Teaching Hospital (UNTH) Enugu, Nigeria to determine the status of the transition from multiple to single stage operative management.
Patients And Method: The clinical records of all children diagnosed and managed at UNTH between January 2008 and December 2022 (15 years) were reviewed. Demographic data collected included age at first presentation, key clinical features, type of HD, operative treatment (single or multi-stage), type of definitive operation, post-op complications, length of hospital stay, and outcome. Each patient's data were collected on a proforma specifically designed for the study. The data were collated and analyzed by SPSS version 25.
Results: The total number of cases analyzed was 44, comprising 35 males and 9 females with M/F ratio of 3.9:1. The age at first presentation ranged from 2 weeks to 10 years with a mean age of 3.8 years. The key symptoms were delayed passage of meconium (91%), chronic constipation (100%) and abdominal distension (80%). Of the 44 cases, 32 (72.7%) were of the short segment type, 7 (15.9%) ultra-short, and 5 (11.4%) long segments. Operative management of the patients was multi-staged with a preceding colostomy in 31 (70%). 13 (30%) patients who presented early without megacolon had the single=stage procedure done without a preceding colostomy. Majority of the patients, 38 (86.4%), had no post operative complications. The length of hospital stay varied from 8 to 35 days; mean of 18.4 days.
Conclusion: Because of late presentation of patients with HD in our sub-region , multi–staged operative treatment continues to dominate the practice. Targeted public awareness campaigns for early presentation of cases and improved access to paediatric surgical care, remain the cornerstone for achieving the desired transition.

Keywords : Hirschsprung's disease, single vs multi–staged operation.

INTRODUCTION

Hirschsprung's Disease (HD) is a chronic functional intestinal obstruction resulting from congenital absence of ganglion cells in the enteric nervous plexuses of the distal bowel (1,2)
The condition was first described by the Danish Physician, Harald Hirschsprung, in 1888 (3)
It took another half a century before the understanding of the pathophysiology of HD became possible, in the early 1940s, It was then appreciated that the proximal dilated, hypertrophic megacolon was not the cause but a consequence of the disease (4,5).
Subsequently in 1948, Swenson & Bill developed the appropriate surgical treatment for the condition (6,7)
The clinical presentation of HD ranges from neonatal intestinal obstruction to chronic constipation with progressive abdominal distension in the older child (8). Diagnosis is mainly by radiographic studies, ano-rectal manometry and histological examination of rectal wall biopsy (8,9). The principle of the Swenson's operative treatment consists of

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excision of the aganglionic and hypoganglionic segments and restoration of bowel continuity by a coloanal anastomosis. Currently the traditional Swenson's abdomino-perineal pull-through operation done in 2 or 3 stages has largely given way in most centers of the developed countries to a one-stage laparoscopic-assisted, or trans-anal, repairs (4,8,9). This is made possible by the fact that majority of cases of HD in developed countries present in the neonatal age, Unfortunately, this is not the case in the underdeveloped and resource poor countries in which a large number of HD cases still present very late with gross abdominal distension and severe megacolon. The single stage operation is unsuitable in most of such cases, and recourse is often made to the traditional multi-stage procedures (10, 11).

STUDY OBJECTIVE

We retrospectively reviewed paediatric patients managed for HD over a period of 15 years, (2008 to 2022) at the University of Nigeria Teaching Hospital (UNTH), Enugu, South-East Nigeria, to determine the status of the transition from multi to single stage procedure.

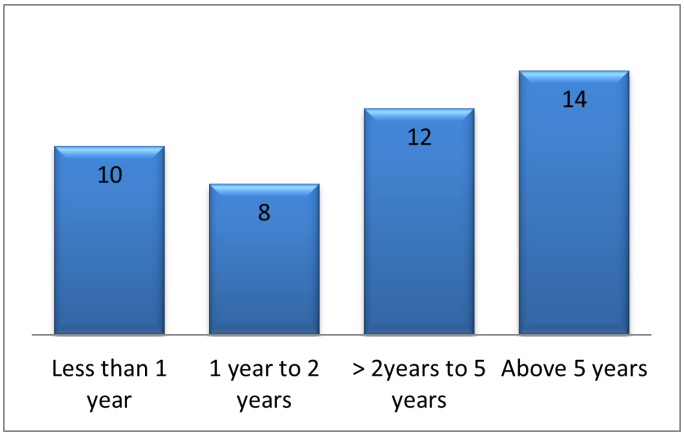
PATIENTS AND METHOD

Clinical records of all children managed for HD between Jan 2008 and Dec 2022 (15 years) were reviewed. Demographic data extracted included sex of the patient, age at 1st presentation, clinical features, method of diagnosis, type of HD, operative treatment (single or multi-stage), definitive operative procedure, post op complications, outcome, length of hospital stay. Data on each patient was collected using a pro forma designed for the study.

RESULTS

A total of 75 children comprising 61 males and 14 females were managed for HD between Jan 2008 and Dec 2022 at UNTH, Enugu, Nigeria, but only 49 clinical records were accessible for review. In 5 out of the 49 cases, diagnosis of HD could not be histologically confirmed and were therefore excluded from the study. Hence total number of cases studied was 44 comprising 35 males and 9 females, with M/F ratio of 3.9:1. Only 10 out of the 44 patients (22.7%) presented below one year of age. 26 patients (59.1%) presented after 2 years, and 14 patients (31.8%) after 5 years.

Figure 1. Age at Presentation.

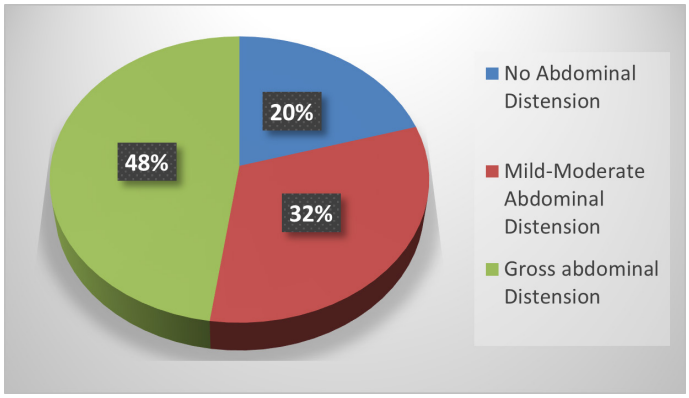


The commonest presenting complaints were chronic constipation, in all the 44 (100%) patients, and delayed passage of meconium in 40 patients (91%). There was significant or gross abdominal distension in 35 (80%) of the patients, while only 9 (20%) patients presented with no abdominal distension.

Table 1. Clinical features in 44 HD patients managed at UNTH Enugu.

Clinical features	No of patients	Percentage
Delayed passage of meconium	40	91
Chronic constipation	44	100
Presence of abdominal distension	35	80
No abdominal distension	9	20

Figure 2. Clinical features in 44 HD patients managed at UNTH Enugu.



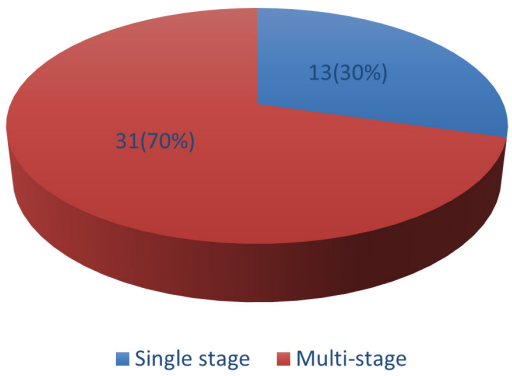
The method of diagnosis was by barium studies in 43 patients (97.7%) i.e. enema or distal colostography; and rectal biopsy for histology in all 44 patients. Only three varieties of HD were seen among the 44 patients managed within the period, namely, short segment in 32 (73%), ultra-short segment in 7 (16%), and long segment in 5 (11%). 4 of the 44 patients had associated anomalies, namely Down's syndrome in 3 (7%) and cardiac malformation (VSD) in 1 (2%).

Table 2. Types of HD seen in 44 patients managed at UNTH Enugu.

Type of HD	No of patients	Percentage
Short segment	32	73
Long segment	5	11
Ultra short	7	16
Total	44	100

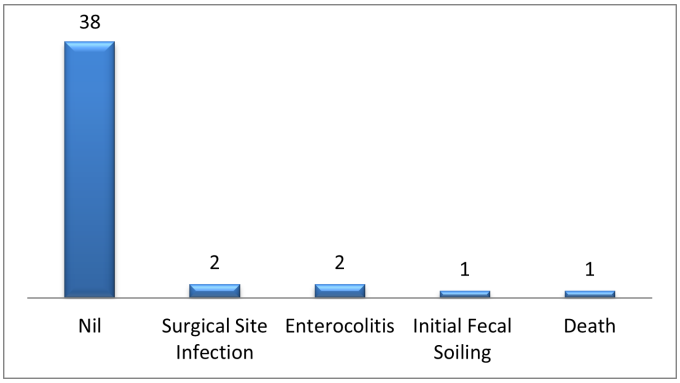
Majority of the patients, 31 (70.4%), had multi-stage procedures. This consisted of an initial colostomy, a definitive modified Swenson's abdomino-perineal pull through procedure, and colostomy closure done during the definitive procedure (2 staged) or later (3 staged). 13 (30%) out of the 44 patients had a one stage procedure namely, modified Swenson's abdomino-perineal pull through in 6 (14%), trans-anal pull through procedure in 3 (7%), and posterior myectomy for ultra-short segment in 4 (9%).

Figure 3. Operative treatment in 44 patients managed for HD in UNTH Enugu.



6 (13.4%) out of the 44 patients had post operative complications, namely surgical site infection in 1 (2.3%), fecal soiling in 2 (4.5%), enterocolitis in 2 (4.5%) and death in 1(2.3%) patient. Majority of the patients, 38 (86.4%) had no post operative complications.

Figure 4. Post–operative complications in 44 patients managed for HD in UNTH Enugu.



DISCUSSION

Hirschsprung's disease is a motor disorder of the colon, caused by the failure of neural crest cells, the precursors of enteric ganglion cells, to migrate completely during intestinal development. The resulting distal aganglionic segment of the colon fails to relax, causing a functional obstruction. The most popular theory of the cause of HD is that there is arrest in the cranio-caudal migration of neuroblasts originating from the neural crest of the developing embryo. The cells fail to reach the distal colon rendering the segment aganglionic, with failure of relaxation leading to functional obstruction. Defects in the differentiation and maturation of neuroblasts into ganglion cells, as well as autoimmune destruction of neuroblasts within the intestine may also contribute to the disorder (12).

HD occurs in approximately1 in 5000 live births, with an overall male/female ratio of 3:1 to 4:1(1, 2, 3). The sex ratio decreases with more extensive aganglionosis, approaching 1:1 for total colonic aganglionosis.

Rectal biopsy for histopathological examination is the gold standard for diagnosis. Absence of ganglion cells in the bowel wall with hypertrophy of nerve endings, and increased acetyl cholinesterase staining are typical findings confirming HD.

Surgery is the mainstay of treatment and the modified Swenson's abdomino-perineal pull through operation done in 2 or 3 stages has largely given way in most centers in developed countries to a one stage laparoscopic-assisted or trans-anal repairs. The added benefits of one stage over multi-stage procedures include earlier resumption of full feeds, shorter hospital stay, and less conspicuous scars (13, 14). The focus of this study is on the transition from multi- to single stage operative management of HD. The study has

revealed a slow transition (30% done single stage), with majority of cases (70%) still requiring multi-stage approach. This is largely due to late presentation as seen in this study, with only 10(22.7%) patients presenting before 1 year of age, while the rest 34(87.3%) presented after one year, and even as much as 14(31.8%) presenting after 5 years of age. In developed countries, majority of HD cases present in the neonatal period with no significant abdominal distention or megacolon. This study has revealed that as much as 35(80%) of the patients presented with significant abdominal distension and megacolon making them unsuitable for a single stage operative procedure. Single stage management of HD has become routine in the developed countries such as Europe and North America (9). In addition to early presentation of cases, there are well established health insurance policies that eliminate out-of-pocket payment barriers for accessing health care.

The other findings in the study are consistent with what is available in the literature (11,15,16,17,19)

Within the sub-region and other underdeveloped and resource poor settings a number of studies have been done in various centers on various aspects of the management of HD (15,16,17,18,19,20,21), but none has focused on the subject of the transition from multi- to single-stage management.

The post operative complications in this study are quite low. When patients are carefully selected for single versus multi-stage procedures, complications are bound to be low.

In a retrospective review of 33 cases in Ile Ife, South-west Nigeria the authors reported 54.5% single stage Swenson's pull through procedure, but with relatively higher rate of complications than the findings in this study (15)

CONCLUSION

On account of late presentation of HD patients with severe megacolon, transition to single-stage operative management remains slow & multi-stage approach continues to dominate the practice in our subregion and most underdeveloped and resource poor countries.

Intensive and targeted awareness campaign, establishment and enforcement of appropriate health insurance policies, improved socio-economic condition of the populace, & improved access to specialist paediatric surgical care, are the essential recipe for achieving and sustaining the desired transition from multiple-to-singe stage procedure in the management of HD in developing and resource poor countries.

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