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A survey of current practices in management of Hirschsprung's disease in Nigeria

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ABSTRACT

Background: Although there are several modalities of treatment for Hirschsprung's disease (HD), there are presently no clear guidelines on treatment of the condition by paediatric surgeons in Nigeria. This survey determines the current approach to treatment among Nigerian paediatric surgeons and should help in establishing a consensus and guidelines for care in this and similar setting. **Materials and Methods:** An online questionnaire was designed using survey Monkey® to determine current clinical and operative management of patients with HD by consultant paediatric surgeons practicing in the Nigeria. The paediatric surgeons were notified by E-mail, which included a link to the survey on survey Monkey®. The survey was also administered at the 12th annual meeting of Association of Paediatric Surgeons of Nigeria in September, 2013, to capture those who did not complete the online survey. Thirty-one paediatric surgeons from 21 different tertiary paediatric surgery centres completed the survey. **Results:** Sixteen (52%) respondents see up to 20% of their patients with HD in the neonatal period. Twenty-six (84%) respondents do routine barium enema. Twenty six (84%) respondents do full thickness rectal biopsy under general anaesthesia (GA). There was no consistency in operative techniques, with transabominal Swenson's operation being practiced by 17 (57%) respondents and 11 (37%) transanal endorectal pull through. 14 (45%) do pull through at any age. 12 (39%) respondents do more than half of their patient as primary pull through. **Conclusion:** Full thickness rectal biopsy under GA is still the vogue with variations in the surgical technique for management of Hirshsprung's disease in Nigeria. Primary pull through procedures is becoming increasingly popular. There's a need for Paediatric Surgeons in Nigeria to come up with a guideline on management of HD, to guide trainees and other surgeons in the care of these patients.

Key words: Hirschsprung's disease, management, primary pull through, rectal biopsy

INTRODUCTION

Hirschsprung's disease (HD) is a relatively common cause of intestinal obstruction in the new-born. It is characterized by the absence of ganglion cells in the distal bowel and extending proximally for varying distances.[1] The first surgical treatment described for HD was a diverting colostomy. Although successful in relieving symptoms and saving the life of the affected child, it was not a satisfactory permanent solution.[2]

Significant advances have been made in the diagnosis and treatment of this condition. These were results of greater awareness of HD, improved neonatal nursing care with specialised neonatal intensive care units, the use of suction rectal biopsy and a larger number of paediatric pathologists who have all contributed to earlier diagnosis and management for these infants. This has resulted in increasing interest in performing a one-stage repair for HD over the last several decades.[2] The introduction of both laparoscopic surgery and transanal endorectal pull through has revolutionised the operative procedures related to the treatment of HD.[3,4]

In spite of these advances, there are no clear guidelines on the way the condition is treated by paediatric surgeons in Nigeria. This survey aims to determine the current approach to treatment of HD among Nigerian paediatric surgeons and should help in establishing a consensus and guidelines for care in this and similar setting.

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MATERIALS AND METHODS

An online questionnaire was designed on survey Monkey® to determine current clinical and operative management of patients with HD by consultant paediatric surgeons practicing in Nigeria. Information requested included location of practice, average number of patients managed per year, proportion that were neonates, modalities for diagnosis, indication for colostomy and preferred type of colostomy. Respondents were also asked the age at which they would consider performing definitive surgery in a well-child and type of pull through. Only 13 paediatric surgeons completed the online survey. The survey questionnaire was sent to paediatric surgeons and due to low response, the questionnaire was also administered to 21 Paediatric surgeons who had not completed the online survey, at the 12th annual meeting of the association between 23th and 28th September, 2013. Eighteen (18) of the 21 paediatric surgeons completed and returned the questionnaire. Participation was completely voluntary. All information collected for this study was confidential. Analysis was done using SPSS, version 15, (SPSS, Inc., Chicago, IL) and online through the survey Monkey® Software.

RESULTS

During the period of the survey, there were 62 paediatric surgeons in 24 paediatric surgical centres in Nigeria. Overall, 31 paediatric surgeons in Nigeria participated in the study [50% response rate Table 1]. The survey indicated that 16 (52%) of surgeons managed an average of 10-15 new cases of HD yearly, [Table 2]. Sixteen (52%) of the respondents see up to 20% of the cases in the neonatal period [Table 2]. Twenty-six (84%) of the respondents do routine barium enema. Twenty-six (84.0%) of the respondents do full thickness rectal biopsy under general anaesthesia (GA) for diagnosis, 3 (10%) respondents do modified suction (grasp and cut) and 2 (7%) surgeons do traditional suction biopsy. Indication for colostomy include absolute intestinal obstruction (18 respondents; 58%), Hirschsprung's associated enterocolities (14 respondents; 45%) and presentation after 5 years (2 respondents; 7%).

There was wide variation in the type of colostomy used; levelling colostomy above transition zone 9 (29%) respondents, loop transverse/sigmoid 9 (29%), devine/defunctioning colostomy 6 (19%), divided sigmoid colostomy 4 (13%), and one of the surgeons had never used colostomy before. There was no consistency in operative techniques, with transabominal Swenson's being practiced by 17 (57%) respondents; 11 (37%) transanal endorectal pull through; 8 (27%) transabdominal endorectal pull through; 6 (20%) surgeons practiced transanal Swenson's, 3 (10%) Duhamel's procedure and only one each do Rehbein and laparoscopy assisted techniques. Forty-five per cent ($n = 14$) surgeons do pull through at any age while 7 (23%) surgeons do pull through at 1 year of age. Twelve of the respondents do more than half of their patient as primary pull through, [Table 3].

Table 1: Location of practice of respondents

Type of practice ($n = 31$)	Number
Teaching hospital	25
Federal medical centre	3
General hospital	1
Not indicated	2

Table 2: Epidemiological characteristic

Average number of patient seen per year	
Number of patient per year	No. of respondents (%)
<10	6 (19.4)
10-15	16 (51.6)
16-20	3 (9.7)
21-25	1 (3.2)
>25	5 (16.1)
Proportion of patient presenting in neonatal period (n=30)	
Proportion of neonate %	No. of respondents (%)
<10	8 (26.7)
10-20	8 (26.7)
21-30	1 (3.3)
31-40	2 (6.7)
41-50	1 (3.3)
51-60	3 (10)
61-70	4 (13.3)
>70	3 (10)

Table 3: Operative technique

Prefer age at pull through	No. of respondents (%)
Any age	14 (45.2)
At 1 year of age	7 (22.6)
At 6 months	6 (19.4)
6 months after colostomy	3 (9.7)
Neonatal period	1 (3.2)
Proportion of primary pull through (n=30)	
Proportion of patient %	No. of respondents (%)
<10	8 (26.7)
>75	7 (23.3)
51-75	5 (16.7)
25-50	5 (16.7)
10-24	3 (10)
Always	1 (3.3)
None	1 (3.3)

DISCUSSION

The presentation of HD varies in relation to age and extent of the disease. In the developed countries, majority of HD cases (90%) are diagnosed in the neonatal period.[5] The challenges in the developing world is delayed presentation with cases presenting late in childhood, adolescence or even in adults.[6,7]

This survey revealed that only about 20% of the HD is seen by the respondents in the neonatal period. The reasons for the delayed presentation in one report included illiteracy, ignorance, inadequate access to specialised centres, poverty and long distances to travel for medical help.[8] With the recent developments and advances, most children with HD should be diagnosed in the neonatal period or within few months after birth.

The definitive surgical management of HD depends on an unambiguous histological diagnosis of bowel aganglionosis. Full thickness rectal biopsy has been the gold standard of confirming the diagnosis of HD. This however requires GA. Majority of the surgeons in this survey still do full thickness rectal biopsy under GA. This is in contrast to an Australian series where rectal suction biopsy (RSB) was done for all 126 children managed over 4 years, with 101 positive results.[5] RSB is a reliable and established procedure for the diagnosis of HD. It is routinely performed in paediatric surgical clinics. It is considered as safe and painless, although a few complications have been described.[9] Inadequacy (in depth, features, and level of the biopsy) still represents an important complication of RSB, and it is the most frequently encountered.[9] In a recent international symposium on HD, the unanimous opinion of a group of experts was that RSB is the current gold standard in the diagnosis of HD.[10] Accuracy of reporting RSB depends on the experience of pathologist, which can be gotten with time. The tool for suction biopsy is not available in Nigeria perhaps because of resource crunch and lack of technology, apart from a shortage of experienced pathologists. The modified suction practice in some centres is welcome. Other centres could collaborate with those centres and the pathologists to make the modified suction biopsy a more frequent practice.

Although barium enema is not essential to confirm the diagnosis of HD; in many cases, it is useful in evaluating the level and extent of aganglionosis and aids in the decision regarding the surgical approach.[10] When the radiological transition zone is situated between the rectum and the sigmoid, it is very likely that the pull through can be performed completely by transanal approach.[11] In this survey, most of the respondents (84%) do routine pre-operative barium enema. This finding is consistent with international experts recommendation.[10]

Although most paediatric surgeons no longer routinely use a diverting colostomy or ileostomy, it is still an important adjunct to the treatment of HD in appropriately selected patients. Even though, patients presenting with complications of HD such as absolute intestinal obstruction or enterocolitis may respond well to aggressive nonsurgical management with antibiotics and colonic wash out,[12] there are some situations in which a stoma is indicated.[2] This survey shows that 58% of the respondents will do colostomy for patients presenting with intestinal obstruction while 45% will do colostomy for patients with Hirschsprung associated enterocolitis. The care giver needs to be aware of other indication for preliminary stoma in selected patients, including those with severe enterocolitis, pneumoperitoneum in the neonatal period, questionable pathology, massively dilated colon, and in some cases those with total colonic aganglionosis or developmental delay.[2] Some children who present with severe life-threatening enterocolitis, and do not respond quickly to intravenous fluids, decompression, antibiotics and other supportive measures may also benefit from preliminary stoma.[13]

This study has shown that there is a wide variation in the preferred management of a child with HD in Nigeria. Transabdominal Swenson procedure is the most preferred, 17 (57%) respondents followed by transanal Soave chosen by 11 (37%) surgeons. This is probably due to preference and expertise of many pioneer Paediatric surgeons in Nigeria. This report is in contrast with a report by Singh *et al*. which showed that Soave procedure is the most commonly performed operation (65%) by members of the Australian Association of Paediatric Surgeons (AAPS).[5] Similar survey of members of British Association of paediatric surgeons (BAPS) in 2008 showed equal preference for both Soave and Duhamel procedures.[14] Since the introduction of complete transanal pull through for HD by De la Torre-Mondragón and Ortega-Salgado in 1998,[4] many groups have reported on this procedure and have shown it to be a safer option.[15-19] It is a simple approach eliminating the need for abdominal access by either laparoscopy or laparotomy. The present survey shows a trend towards transanal pull through in the management of children with HD. The first to describe the single-stage pull through were So and *et al*. in 1980.[20,21] Since these initial reports, there have been

many additional single and multi-institution studies published that demonstrate the safety of single-stage repairs for HD.[11,17,18,22-24] The risks of contamination and adhesion formation are minimal; the procedure does not damage the pelvic structures, it is not expensive, and it has the best cosmetic results.[25] However, one-stage repair has also been described for all of the commonly performed operations for HD.[4,16,23,26,27] The single-stage repair performed laparoscopically has been reported.[3,28]

This single-stage approach has made it possible to complete the surgical treatment in one hospitalisation; this approach avoids the need for readmission to the hospital and also prevents the stigma of having a colostomy.[2] In developing countries, where access to medical care may be hampered by poverty, poor transportation, and communication systems, the implementation of single-stage repair has made it easier to provide quality care to children with HD.[6,7,29]

A survey of the BAPS in 1998 revealed that 41% of the pull through procedures performed at that time were through primary approach.[30] A recent survey of BAPS members by Bradnock and Walker revealed that 86% of 36 respondents now performed primary pull through in a well-child.[14] Singh *et al*. in a survey of 126 children managed for HD on a prospective basis over 4 years by members of the AAPS reported that 51% of the children had primary pull through.[5] The findings of the of the present survey with only 40% of the respondents performing primary pull through for more than half of their patients is rather low compared with other reports.[5,14] The approach of primary pull through can be upheld by paediatric surgeons practicing in resource constrained setting like ours with overall cost containment for the health system and the patient, who often pay for health services out of pocket.

CONCLUSION

Full thickness rectal biopsy under GA is widely practiced. There is wide variation in the surgical technique for management of Hirshsprung's disease in Nigeria. Transanal pull through and Primary pull through procedures have become increasingly popular. This study is limited by its low response rate. We recommend routine preoperative barium enema to aid choosing appropriate approach. Efforts should be made to increase utilisation of RSB. There are still indications for a preliminary stoma and this should be considered in patient with severe enterocolitis, pneumoperitoneum in the neonatal period, questionable pathology, massively dilated colon in whom initial colonic washout is unsuccessful, and in some cases those with total colonic aganglionosis or developmental delay.

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