Original Article

Hirschisprung's disease: Factors Affecting the Outcome at The National Ribat University Hospital, Khartoum, Sudan, 2007 to 2011

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ABSTRACT

Background: Hirschsprung's disease (HSD) remains the most frequent cause of child intestinal obstruction.

Objectives: to evaluate the effect of different factors at the final outcome, postoperative complications and the hospital stay of children with HSD.

Methodology: It is a retro-prospective analytical observational hospital based study, involving all the cases presented initially to the Paediatric Surgery Centre at the National Ribat University Hospital and confirmed to be a case of HSD. The data were collected using a questionnaire and analyzed by the software SPSS version 17.

Results: Sixty four patients were involved in this study with male to female ratio of 5:1. The mean age at the first time of presentation was 9 days and the bulk of the patients seen from the center of Sudan. The emergency presentation accounted for 21.9% of the cases with the delayed passage of meconium and constipation as the main presenting symptoms.

Complications occur mainly at day 13 post operatively with colostomy prolapse as the commonest type of complications. The age of the patients at the time of Pull Through Procedure (PTP) was ranging between 7 to 72 months and the mean body weight found to be 11.94 kg. Complications following PTP occur in 18.8% of the cases, with wound infection accounting for 15.6% of the cases. The total duration of hospital stay post operatively after reversal of colostomy was found to range between 6 and 60 days with a mean of 8.77 days and SD of ± 6.657.

Conclusion: The average hospital frequency of HSD in our study is compared to that encountered in European countries and Northern American countries. Early diagnosis and treatment is essential for better outcome. Emergency presentation, age and the weight at Pull Through Operation and at the time of closure of colostomy, significantly affect the outcome and prolong the hospital stay.

Key words: Hirschsprung's Disease, colostomy, Pull Through Operation.

Among congenital digestive tract malformations, Hirschsprung's disease (HSD) remains the most frequent cause of child intestinal obstruction[1]. Hirschsprung's disease is a functional obstruction due to lack of ganglion cells in the distal colon.

Harald Hirschsprung, a Danish physician, wrote early texts about the disease[2]. In the developed countries good knowledge of the disease allows for an early diagnosis and care, but in developing countries the diagnosis is often late largely due to the use of traditional enema, the inaccessibility or the non-availability of some clinics. This study is aiming to evaluate the effect of different factors at the final outcome, postoperative complications and hospital stay.

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MATERIALS AND METHODS:

This is a retrospective analytical observational hospital based study of HSD carried out in the Paediatric Surgery Center at The National Ribat University Hospital (PSC/RUH) in Khartoum, a reference hospital in Sudan that receives patients from Khartoum and the rest of the country, in the period from January 2007 and December 2011. The study involved

all the cases presented initially to the PSC/RUH and confirmed to have HSD. Patients were included in the study when their first presentation was before any intervention and the diagnosis of HSD confirmed by biopsy and initial treatment and definite treatment and follow up carried out in RUH. The study was approved by the institutional ethical committee for medical research. Data collected included variables related to clinical and therapeutic information together with the information related to hospital stay and complications. Data were collected, retrieved and analyzed by the software SPSS version 17. To determine the statistical significance of differences, the Pearson test was used and probability test (P. value) with P value less than 0.05 considered as significant.

RESULTS:

Sixty four patients were reviewed, the mean age at presentation was 9.02 days (range 1 to 60 days), male to female ratio was 5:1 and 60.4% of them were from the center of Sudan. 14 patients (21.9%) presented as emergency cases. Delayed passage of meconium and constipation recorded as 87.5% and 79.7% respectively. Fever was recorded in 7.8%, abdominal distension was found in 40.6%, vomiting in 15% and no patient presented with any associated clinically obvious congenital anomaly. Barium enema was done in 32.8% of cases and the site of the collapsed segment of the colon showed in (figure1). No reported cases in the study of ascending colon or total colonic involvement. The mean width of dilatation was 16.32mm with a range of 12 to 20 mm and SD of ± 2.023 in the study group. The initial treatment done was defunctioning colostomy then biopsy in 35.9% of the cases, conservative treatment then rectal biopsy in 29% of the cases and leveling colostomy with the associated biopsy in 34.4% of the cases (Figure 2).

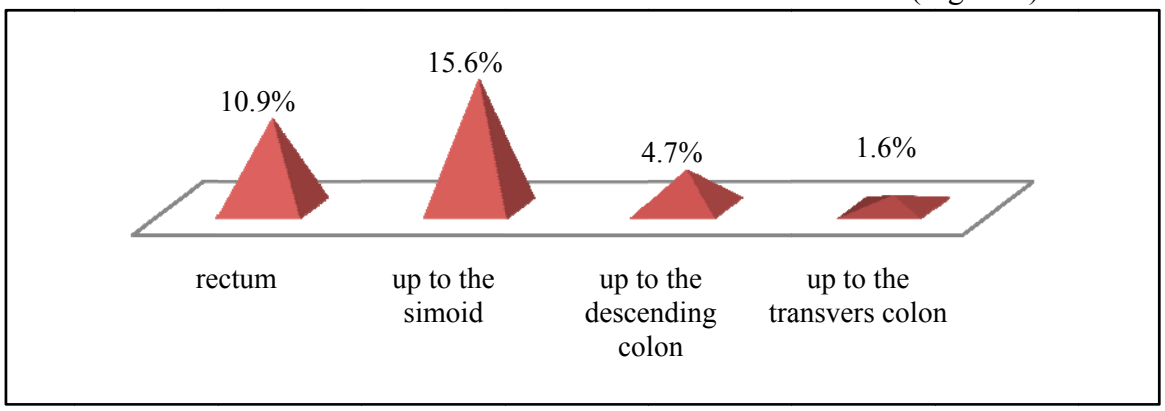

Figure 1: Collapsed segment of the colon in barium enema done for the studied patients.

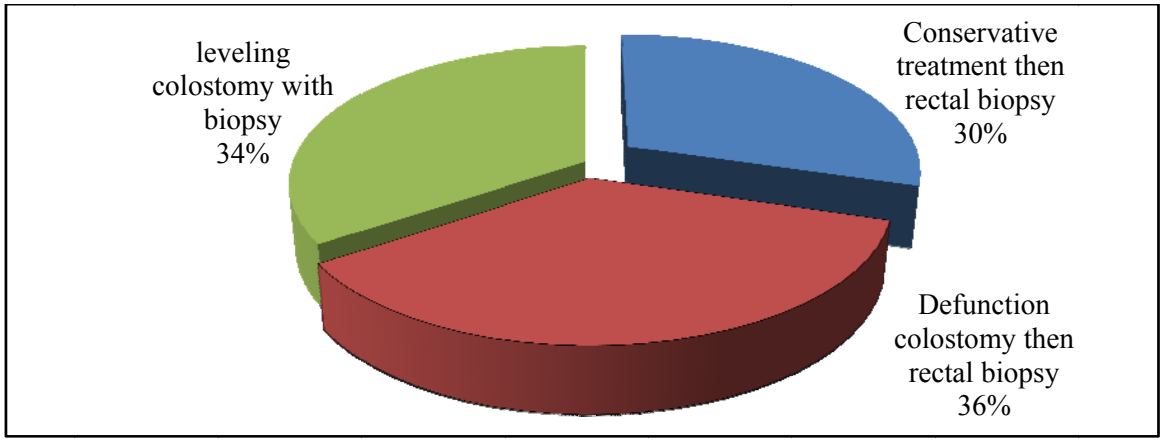

Figure 2: The initial surgical treatment done for the studied patients.

Post-operative complications after the initial surgical treatment were observed in 10.9% of the cases and it occurs mainly at day 13 post operatively. All complications occurred between day 7 and day 30 post operatively.

The most common type of complication observed was colostomy prolapsed in 6.3% followed by necrotizing enterocolitis in 3.1% and skin excoriation in 1.6% of the cases. These complications were corrected surgically in 7.8% of the cases and conservatively in 3.1%.

The age of the patients at the time of the Pull Through Procedure (PTP) ranged between 7 and 72 months with the mean of 30.36 months and SD of ± 11.798. The weight of the patients also observed to be between 7 and 21 kg with a mean of 11.94 kg and SD of ± 1.902.

Post-operative complications after PTP occurred in 18.8% which was mainly in the form of superficial wound infection in 15.6% of cases. The complications occurred between day 2 to day 8 post operatively with a mean of 3.25 day and SD of ± 1.685. The duration of total hospital stay after Pull Through operation was found to be between 10 and 30 days with a mean of 14.61 days and SD of ± 3.240.

At the time of closure of colostomy after PTP, the mean age of the patients was found to be 35.72 months (range between 9 to 84 months (SD of ± 12.606); while the patient's weight was found to range between 8 and 22 kg with mean weight of 12.87 kg with SD of +/- 2.178.Post-operative complication occurred only in 6.3% of the cases. The total duration of hospital stay post operatively after reversal of colostomy was found to range between 6 and 60 days with a mean of 8.77 days and SD of ± 6.657.

The last follow up for all patients was between 2 and 21 weeks with a mean of 10.4 weeks post operatively. Long term post-operative complications were found only in 1.6% of the cases.

DISCUSSION:

The average hospital frequency in this study is 12.8 cases per year, as compared to that encountered in European countries [3] and in Northern American countries, [4,5] may suggest that HD is frequent in Sudan. However, this frequency seems to be under-assessed since this does not take patients who died before they come to the medical care or those with hidden, non-obstructive forms and also patients that were excluded from the study because of receiving initial treatment outside our center.

The male predominance in this series is comparable with other series[6]. The average age at diagnosis was far higher than reported in series from the developed countries, where diagnosis is almost made right at birth[3,5,7]. This delay in terms of diagnosis and treatment is due to some practices that prevail in our context. Due to some cultural reasons, evacuation enemas are systematically practiced on children, leading to a delay in the diagnosis of patients with delayed passage of meconium or chronic constipation. Patients are referred to hospital only in cases of complications.

Radiographic examination together with clinical data allowed diagnosis in 39.1% of patients in comparison with a study done in Burkina Faso by E Bandré et al, who did it in 13% [8]. Opaque enema examination was necessary for the diagnosis of HD by showing the narrow area, the transitional cone and the broadened area; the opaque enema is obviously a necessary examination for the diagnosis, it was done for 32.8% of patients, but it does not always show the exact extent of the aganglionic part[9,10]. The histological study of rectal biopsy, which is the diagnostic confirmation of the disease, was done for every patients.

Colostomy was made in 70% of patients (whether leveling or defunctioning followed by biopsy). This result is higher than of the others [8,11]. The current trend is surgical operation as soon as possible without any colostomy, except in long segment, or total colonic aganglionosis or complicated cases of HD or the cases seen late with big bowel[3,6,11].

The average waiting time before the final treatment was long (9 to 84 months mean 35.72 months) compared to those reported in

literature[11]. This could be explained by the need for a good preparation of patients to final surgical operation (treatment of cachexia, anaemia, etc). The average age at the surgical operation was three years, quite higher than that mentioned by European and Northern American authors where surgery is practiced just few days after birth [3,6,7].

Post-operative complications after PTP occur in 18.8% which is better than that of Ekenze SO [12] who stated that 48.8% of his patients experienced at least one postoperative complication. The duration of total hospital stay after the pull through operation was found to be between 10 and 30 days with the mean of 14.61 days and SD of ± 3.240.

CONCLUSION:

The average hospital frequency of HSD in this study is compared to that encountered in European countries and Northern American countries. Early diagnosis and treatment is essential for better outcome. Emergency presentation, age and the weight at Pull Through Operation and at the time of closure of colostomy, significantly affect the outcome and prolong the hospital stay.

COMPETING INTERESTS:

The authors reported no conflict of interest and no funding was received for this work.

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