Hirschsprung's disease: Management problem in a developing country

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ABSTRACT

Background: The management of Hirschsprung's disease remains a problem in developing countries. Our aim is to identify the main epidemiological, clinical, and therapeutic characteristics of Hirschsprung's disease at the University Child Hospital Charles De Gaulle of Ouagadougou (CHUP-CDG). **Patients and Method:** It is a retrospective study carried out in the period from January 2001 to December 2007 in the Surgery Unit at CHUP-CDG, which is a reference centre for Paediatric Surgery in Burkina Faso. **Results:** There were 52 patients (M: F=3.3:1). The annual incidence was seven cases. Age at presentation and diagnosis ranged from two days 10 years (median 20 months). Twenty five patients were from poor socio-economic conditions. Presentations were mainly intestinal obstruction, chronic constipation and enterocolitis. There were two cases of associated trisomy 21. Average age at operative intervention was 3.17 months. The rectosigmoidal form was the most frequently encountered. Over two-thirds (67.31%), with no complications at presentation, had benefited from nursing before their final treatment. A temporary colostomy was requested in case of complication. Swenson's technique was practiced for all the patients who underwent surgery operation. The assessment of functional results in eight patients after an average decrease of 3.5 years gave excellent results. Post-surgery complications were mainly enterocolitis in 12% of patients. Mortality rate was 16%. **Conclusion:** Management of Hirschsprung's disease is a problem in Burkina Faso. It is characterised by its late presentation and difficult diagnosis due to inaccessibility and the non-availability of some investigation services (barium enema, histochemistry, and histology), resulting in high morbidity and mortality rates. Effective technical capacities, adequate staff training, and public education will be necessary to improve care quality.

Key words: Clinic, evolution, Hirschsprung's disease, therapeutic

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INTRODUCTION

Among congenital digestive track malformations, Hirschsprung's disease (HD) remains the most frequent cause of child intestinal obstruction.[1] 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. Our study aims at determining the main epidemiological, clinical, and therapeutic characteristics of HD in the University Child Hospital Charles De Gaulle of Ouagadougou (CHUP-CDG) in Burkina Faso, to highlight some particularities of this disease in a developing country.

PATIENTS AND METHODS

This is a retrospective study of HD carried out in the Paediatric Surgery Unit at CHUP-CDG of Ouagadougou, a reference hospital in Burkina Faso that receives patients from Ouagadougou and the rest of the country, in the period from January 2001 and December 2007. Located in the heart of West Africa, Burkina Faso is notably an agricultural country with a surface area of 274,000 square kilometres. Its population is estimated at 13,902,972 in 2006. Half of this population is under -15 years of age. Data collected included variables relating to epidemiological, clinical and therapeutic information. Functional results after a surgery were classified into three categories, namely excellent, average and bad [Table 1].

RESULTS

Fifty two HD cases were collected over the seven-year period, accounting for about seven cases per year. Over

Table 1: Functional result score

Score	Criteria
Excellent	Soft stools once a day
Average	Constipation controlled by drug
Bad	Constipation not controlled by drug, soiling, incontinence

the same period, 6,329 paediatric cases were registered in the unit. HD accounted for 0.82% of the cases. There were 40 (77%) boys and 12 (23%) girls (M; F= 3.3:1).

Patients ages ranged between two days and 10 years (median age: 20 months). Nineteen (36.5%) of the patients were newborn, 21 (40.4%) were infants and 12 were (23.1%) children. Twenty five patients were from poor socio-economic conditions. Clinical reasons that allowed suspecting the diagnosis of HD are summarised in Table 2.

All patients underwent radiography in an unprepared abdomen, and this revealed a gaseous distension (n = 21), a lack of rectal aeration in newborns (n = 19), a stercoral stasis (n = 7) and gas-fluid levels (n = 5). Barium enema conducted in 23 patients (42%) highlighted a disparity in terms of size and transitional area, all in recto sigmoid. Rectal biopsy confirmed the diagnosis in 18 patients. Trisomy 21 was suspected in two patients.

Medical treatment was instituted on the admission in 13 patients (25%) and consisted of the treatment of both hydro electrolytic disorders and anaemia (blood transfusion) and antibiotic therapy. Twenty seven patients received nursing as waiting therapy. A sigmoid colostomy cited in the left iliac fossa was made in 25 patients (48%); eight had intestinal obstruction, six-enterocolitises, and 11 others with no complications. Nine of our patients had primary pull-through without colostomies, but none had endoanal pull-through (De la torre's technique).

The average duration of the colostomy was 3.86 months (range: 32 to 240 days). Twenty five patients with sigmoid colostomy benefited from definitive treatment (pull-through) and 23 absconded from further treatment. Among those who received the final treatment, 18 patients had their colostomies closed. The average duration between the diagnosis and the final operation was 3.5 months. The average age for the final surgery was 24 months (range: 12 to 125 months).

Seven patients had complications, including abscess, enterocolitis (six) and prolapse (one). Four patients died in the postoperative period, accounting for a mortality rate of 16%: one patient died from acute lung oedema while three died from enterocolitis on the day 10, 12 and 25 respectively. After an average four-year follow-up (range: two months to seven years), the functional results were excellent in eight patients, as per our score.

Table 2: Clinical presentation

Clinical features	Number	%
Chronic constipation + positive rectal test	22	42
Acute intestinal obstruction + chronic medical history of constipation	5	10
Neonatal intestinal obstruction	19	36
Enterocolitis	6	12

DISCUSSION

The low hospital prevalence in our study (7.4 cases per year), as compared to that encountered in European countries[2] and in Northern American countries,[3,4] may suggest that HD is less frequent in Burkina Faso. However, this prevalence seems to be under-assessed since this does not take patients who died before their care or those with hidden, non-obstructive forms using daily traditional enemas into account.

The male predominance in our series as compares with other series.[4] The average age at diagnosis was far higher than reported in series from the developed countries, where diagnosis is almost made right at birth.[2,5,6] 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 passage of meconium or chronic constipation. Patients are referred to hospital only in cases of complications.

Radiography examination in unprepared abdomen together with clinical data allowed diagnosis in 9 patients. Opaque enema examination was necessary for the diagnosis of HD by showing the narrow area, the transitional cone and the broadened area; this was not always possible because of a limited technical capacity in our radiology unit, and the high costs of radiological examinations. The opaque enema is obviously a necessary examination for the diagnosis, but it does not always show the exact extent of the aganglionic part.[7-8] The histological study of rectal biopsy, which is the diagnostic confirmation of the disease, was unfortunately done only in 18 patients (34%) because of the limited technical capacity and the high cost of this examination for parents who were in majority from poor socio economic.

Colostomy was made in 25 patients (48%). This result is higher than Suita's[9] that reported a rate lower than 35.8%. The current trend consists in surgical operation as soon as possible without any colostomy, except in long segment, or total colonic aganglinosis or complicated cases of HD or the cases seen late with

big bowel.[2,5,9] Non-compliance with nursing obliged us to deliberately offer colostomy in patients from poor socio-economic conditions as soon as possible so as to avoid iatrogenic complications due to traditional practices. We routinely offer colostomy in this group of people because of their non-compliance to nursing which comprises the use of rectal catheter, lubricant or physiological serum. This nursing is considered too constraining or expensive by the parents; hence they regularly resort to traditional healers and some cultural practices like the use of traditional enema, using "pear with decoction," a source of deadly complications such as intestinal perforations, intestinal necroses.[10]

The average waiting time before the final treatment was long compared to those reported in literature.[9] This could be explained by the need for a good preparation of patients to final surgical operation (treatment of cachexia, anaemia, etc). Only 25 patients had definitive surgical treatment, this is lower than that reported by Singh (64.57%).[4] This could be explained by the high number of patients that absconded (23 patients). Could they have died from enterocolitis, other complication or of related undiagnosed malformations? The average age during the surgery operation was three years, quite higher than that mentioned by European and Northern American authors where surgery is practiced just few days after birth.[2,5,6]

Apart from enterocolitis, which remains one the main causes of death in most of reported series,[10] acute lung oedema, probably due to co-existing undiagnoses-associated malformation or hydro-electrolytic overload due to lack of medical surveillance, is another factor to watch out for.

In conclusion, despite its low hospital prevalence, HD raises some difficulties in terms of care at CHUP-CDG. Its diagnosis is most often late and difficult due to inaccessibility or the non-availability of some supportive services (barium enema, histochemistry, histology). Non-compliance with nursing is an indication for immediate colostomy in patients from poor socio-economic conditions so as to avoid iatrogenic complications due to traditional enema. The acquisition of an efficient technical support centre (radiography, pathological anatomy with a possibility to make extemporaneous analysis of biopsies to clarify the quality of the colon to reduce), together with sensitisation and education of our people on their health issues, will improve the quality of care of this disease in Burkina Faso.

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