Hirschsprung's Disease in Bahrain

K.H. Al-Ras Romani*, K. Zaber**
Mannan Khan***

INTRODUCTION

IN 1861, the great Danish Paediatrician, Hirschsprung, gave a lecture to Berlin Paedatric Society describing the clinical and autopsy findings in two infants who had died of constipation associated with dilatation and hypertophy of the colon. Since then the Paediatricians are more aware of this fatal disease.

In this paper, we present our experience with Hirschsprung's disease in Salmaniya Medical Centre since May 1980, when the Paediatric Surgical Unit was established. Over the last sixteen months we detected ten cases of Hirschsprung's disease. The incidence of this condition is fairly high in Bahrain. A prospective study was carried out in Salmaniya Medical Centre to study the incidence, clinical presentation, radiological & pathlogical findings and the result of the surgical treatment in Hirschsprung's Disease.

MATERIALS AND METHODS

Ten cases of Hirschsprung's disease were detected over the last sixteen months from May 1980 to September 1981. This is relatively a large number for a small island with 350,000 population and 10,000 births/year. Six of these patients were neonate, presented at the first week of life. Two were two years old, one was eight years and one was ten years. (Eight of these patients were Bahrainis and two were Pakistanis). The main clinical presentation of the neonates were the delay in passing meconium, lazy bowel motion and mild abdominal distension. One baby had bile stained vomiting and presented with meconium plug syndrome. The other four children presented with chronic constipation since birth and occasional abdominal distension. Six were males and four females. None had a family history of Hirscsprung's disease. In neonates, barium enema showed variable colonic distension above a normal or narrowed rectum and colon. Repeated plain abdominal x-ray after 24 hours showed barium residue. In the other children the barium enema showed a hugely dilated colon above a mildly narrowed rectum. The diagnosis was confirmed by complete rectal biopsy, two cms. above dentate line. This showed absence of ganglion and hypertrophic nerve fibres. Eight of our patients had aganglionosis of the rectum and recto-sigmoid, but in two cases the aganglionic segment was extending up to the descending colon.

Three of the neonate had defunctioning loop colostomy, after the diagnosis was confirmed by rectal biopsy. The parents of the other three refused any surgical intervention. Endo-rectal pull through performed in five cases. Two were one year old at the time of endo-rectal pull through, one was eight and one was ten years old. One of the neonate who was premature, failed to thrive after colostomy, inspite of presence of normal ganglions distal to the colostomy. This patient was admitted in many occasions for management of the inflamed skin around the colostomy. The mother was careless and did not show any interest in her baby. We had no alternative but to do her definite procedure at five months when she weighed three Kgs. only. Four of our patients had a Soave (9) and one had Boley (3) modification of endo-rectal pull through. The colostomy closed at the time of definite surgery in two patients, and three weeks after in one patient. The two older children had no clostomy.

* Consultant,
Pediatric Surgeon,
Salmaniya Medical Centre,
State of Bahrain.

** Senior Resident,
Salmaniya Medical Centre,
State of Bahrain.

*** Chief Resident,
Salmaniya Medical Centre,
State of Bahrain.

RESULTS

All these five patients, had full post operative recovery. There was no anastomotic leak or wound infection or faecal fistula. The mean follow up was nine months. None had anal stenosis or faecal incontinence. One infant developed necrostising entercolitis three months after surgery. This cured with medical treatment. There was no operative or post operative mortality in his group.

DISCUSSION

The incidence of Hirschsprungs disease is variable in the literature. In 1946, Scott (8), reported the incidence of Hirschsprungs disease 1/8000. Three years later, Bodian (2) had an incidence of 1/20000 to 1/30000 births. Burnard (4), in 1950, presented the incidence 1/5000 and Lister (7) 1/10000. In our short experience, we report the incidence in Bahrain, 1/4000. Kleinhaus (6) reported 40% of the cases in his survey, were diagnosed in the first three months of life, and 60% by the end of the first year. But only 15% of these cases were detected in the first month of life. 60% of our cases were diagnosed in the first week of life. The overall

male to female ratio has been reported as ranging from 3 : 1 to 5 : 1 (1).

Kleinhaus (6) reported a male to female ratio of 3 : 8. In patients with family history of Hirschsprungs disease, the male to female ratio has been reported 1 : 1 (1). In our small group the male to female ratio was 3 : 2. None of our patients had a family history of Hirschsprungs disease. But there is a high incidence of intermarriages in Bahrain. The extent of the aganglionic segment did not reveal any significant variation from previous reports. Enrenpreis (5) found Hirschsprungs disease to be confined to the rectum and recto-sigmoid in 77% of his cases. Kleinhaus (6) reported 75% involvement of the rectum and recto-sigmoid. In this series, 80% of the cases had aganglionic rectum and recto-sigmoid and 20% had involvement of the descending colong. Necrotising Enterocolitis was reported as a major cause of death. The mortality is about 30% (7). One of our neonate, in which the colostomy was refused by the parents, had severe necrotizing enterocolitis and did not survive.

The rate of post-operative complication varies with different anastomosis. It was reported 1.1% with Soave, and 5.8% with Boley's procedure. The rate of faecal fistulae is about 1.1% with both techniques and the anal stenosis 9.4% with Boley and 15% with Soave (9). The incidence of faecal incontinence about 2%. None of our patients had any of these complications.

The incidence of post operative necrotizing enterocolitis reported 15% with Soave and 2.1% with Boley's techniques (3). One of our patients presented with necrotising enterocolitis three months after Soave operation. She did well with medical therapy.

Post-operative mortality with endo rectal pull through was reported recently 1% in the survey of the Members of the Surgical Section of the American Academy of Paediatrics (6). None of our patients died as a result of surgery.

Hirschspungs disease is a relatively common disease in neonates. It should be kept in mind, remembered in every infant with lazy bowel motion and each child with chronic constipation. The diagnosis is simple, the surgical treatment is curative and the operative mortality and morbidity are low.

SUMMARY

Ten cases of Hirschsprungs disease presented from Salmaniya Medical Centre. The clinical presentation, the radiological appearance and the pathological findings were described. The surgical treatment and its results were discussed.

REFERENCES

1. Bodian M Carter CO; A Family Study of Hirschsprung's Disease. Ann. Hum. Genet, 26 : 261, 1963.

2. Bodian M Stephens, FD A B C H 1949 "Hirschsprung's Disease" Lancet 1, 6.

3. Boley S J : New Modification of Surgical Treatment of Hirschsprung's Disease. Surgery 56 : 1015, 1964.

4. Burnard E D (1950). H D in Infancy. Br. Med. J 1, 151.

5. Enrenpreis, Th : Hirschsprung Disease (Cuiuario; Year Book Medical Publishers Ing., 1970)

6. Kleinhans S, Scott J. Boely, Michael Sheran, and William K Sieblie (1979). Hirschsprung's Disease A surgery of the Members of the Surgical Section of the American Academy of paediatrics. J Paed. Surg. Vol XIV No. 5 - 589.

7. Lister, J Hirschsprung's Disease (1978) Neonatal Surgery 2nd Edition 443.

8. Scott, W J M and Serenati, OS. (1946) "Megacolon" Mechanism and Choice of Treatment Surgery 20, 603.

9. Soave F : Die Nahtlose Colon — Anastomose Nach Extramucoser Mobilierung Un Herabzienhung Des Recto-Sigmoid Zur Cuirurzischen Behandlung Des M Hirschsprung ZBI Chir 88 : 31, 1963.

10. Swenson O, Sherman J, Fisher J H : Diagnosis of Congenital Megacolon; An Analysis of 501 patients. J Paediatric Surg. 8 : 587, 1973. □□