Archives of Disease in Childhood, 1977, **52**, 713–715

Anorectal myectomy in treatment of ultrashort segment Hirschsprung's disease

Report of 26 cases

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SUMMARY A diagnosis of Hirschsprung's disease should be considered in children with constipation. An accurate neonatal history of bowel function and testing of anorectal pressure responses will aid the diagnosis. In the period 1971–75 inclusive, 140 children, aged 6 months to 14 years, were investigated by anorectal manometry. 26 showed a failed inhibition response to rectal dilatation, suggesting Hirschsprung's disease and were treated by anorectal myectomy. In 24 the disease was confirmed histologically. Two specimens were diagnostically unsuitable. 4 required repeat myectomies, and 3 anterior resection. At follow-up all had normal bowel movements without soiling.

Many children attend hospital because of constipation and soiling, the latter being the main reason for seeking medical help. Outpatient visits to medical paediatric and psychiatric clinics are interspersed with admissions for 'emptying out' procedures and toilet training. These children are labelled as encopretics, but such colourful descriptions as 'potty refusal syndrome' are to be found. Among these unfortunate children there will be a few with Hirschsprung's disease who unless fully investigated will go undiagnosed and untreated (Clayden and Lawson, 1976).

Hirschsprung's disease in the older child usually involves the terminal rectum and is amenable to treatment by anorectal myectomy (Bentley, 1966; Duhamel, 1966; Nissan *et al.*, 1969; Lynn and Van Heerden, 1975). We report the experience of investigating a group of children with constipation and soiling attending the Edinburgh children's hospitals.

Patients and methods

In the period 1971–75 inclusive, 140 children who had suffered chronic constipation from the first year of life were referred for investigation with a view to diagnosing Hirschsprung's disease. 35% had continuous soiling and this included almost all those with the longest period of medical management. Many of this group had been labelled encopretic, but 9 were subsequently found to have Hirschsprung's disease. Among the 9 were two cousins, of whom a sister had had a Swenson rectosigmoidectomy and a brother had died at 13 days with 'faecal impaction'.

A specific note was made of any delay or difficulty in passing meconium or any change in bowel habit with the introduction of solid feeds. Any diarrhoea-like illness was recorded, especially if associated with previous constipation and abdominal distension. Patients with neurological deficits or any congenital anorectal anomaly were excluded.

Barium enema was not part of the investigation, though many patients had had this before referral. It was of limited diagnostic value in the present series. A rectal examination was always performed at the initial visit to assess sphincter tone and faecal loading. Patients with excessive faecal loading and a dilated rectum were put on a regimen of thrice weekly enemas at home, administered by the Paediatric Community Nursing Service. This allowed the rectum to return to a normal calibre before carrying out manometry.

All patients underwent anorectal manometry as an outpatient procedure using the technique described by Lawson and Nixon (1967). 26 patients (21 males and 5 females ranging in age from 6 months to 14 years) showed a failed inhibition response on rectal dilatation, supporting a diagnosis of Hirschsprung's disease (Lawson and Nixon, 1967; Schnaufer *et al.*,

Received 17 February 1977

1967). They were treated by anorectal myectomy. One boy of 14 was known to have Hirschsprung's disease, having had a Swenson operation at 2 years of age; but constipation had continued.

Myectomy technique. All myectomies were performed by the same surgeon (W.G.S.). No special bowel preparation was carried out. Both Bentley (1966) and Nissan *et al.* (1969) recommended washouts and antibiotics both before and after operation. However, a bolus of stool tends to lie at the upper level of the aganglionic segment and is a useful guide to the length of myectomy required. It can be easily packed clear of the operative field.

With the patient in the lithotomy position the anus is stretched and a long-bladed nasal speculum is inserted as a retractor. A curved incision is made at the mucocutaneous junction on the posterior wall of the anal canal and the internal sphincter identified. In the posterior midline a plane is established between mucosa and muscle and a strip 0·5–1 cm wide which includes internal sphincter and rectal muscle is excised proximally for a distance of 5–10 cm. The transverse incision is closed with interrupted catgut. Occasionally the mucosa is breached but can be easily repaired. The excised strip is correctly orientated, marked, and pinned on a strip of cork for pathological examination.

Patients were able to go home on the third postoperative day. A laxative was prescribed for 3 to 4 weeks after operation.

Results

All 26 patients were followed up at regular intervals and the length of review varied from 6 months to 5 years. 21 myectomy specimens had features typical of Hirschsprung's disease with absent ganglion cells and large abnormal nerve bundles. 9 were totally aganglionic, and 12 showed normal ganglion or a transitional zone of scattered ganglia, at the upper end. All these specimens were long enough to extend beyond the distal 3 cm segment which is usually aganglionic.

Of the 9 with totally aganglionic specimens, 2 did not require further procedures and included the boy who had had a previous resection. The length of his myectomy was 6·5 cm and extended to the level of his original anastomosis. 4 had repeat myectomies in order to get above the aganglionic zone. In 2 the myectomy was repeated twice. In the 3 remaining patients biopsies taken through a sigmoidoscope indicated that the aganglionic segment was beyond the scope of myectomy and they were treated by anterior resection. The distal resection was carried down to the upper level of the previous myectomy. The aganglionic segment in these patients was found to extend proximally to the level of the peritoneal reflection or just above it. In the remaining 5 specimens which were less than 5 cm long, 3 showed large nerve bundles and muscle hypertrophy, and 2 had no typical features that were diagnostically valuable.

All patients had temporary soiling postoperatively for 2 to 3 weeks, but at follow-up no patient was found to be soiling. One patient had bleeding which required resuturing of the myectomy site.

17 patients had achieved normal regular bowel movements after the initial myectomy and included the 5 with short myectomy specimens. The remaining 9, after repeat procedures, had normal function at follow-up (Table 1).

Discussion

In 1966 Bentley reported his experience on the treatment of what he described as ultrashort segment Hirschsprung's disease by posterior anorectal myectomy. He defined ultrashort segment as aganglionosis of the distal third of the rectum or less, being thus akin to achalasia of the internal sphincter (Hurst, 1934). Unlike patients with more extensive Hirschsprung's disease, this group do not present with neonatal obstruction. They experience delay and difficulty in passing meconium and continue into childhood with constipation. The age at which the diagnosis of Hirschsprung's disease was made is similar to that found by Nissan and Bar-Maor (1971) and Lynn and van Heerden (1975) (Table 2).

Table 1 *Results in 26 patients relating histology to initial myectomy and additional procedures required*

Histology	*No. of cases*	*Initial myectomy*	*Additional procedure*
Aganglionic + transitional zone	12	Normal bowel movements	
Totally aganglionic	3	Constipated	Anterior resection; normal bowel movements
Totally aganglionic	2	Normal bowel movements	
Totally aganglionic	4	Constipated	Repeat myectomies; normal bowel movements
Large nerve bundles + muscle hypertrophy	3	Normal bowel movements	
Not diagnostic	2	Normal bowel movements	

Table 2 *Age at time of diagnosis of Hirschsprung's disease in present and other series*

	Age at time of diagnosis		
	0–1 m	*1–18 m*	18 m–14 yr
Nissan and Bar-Maor (1971) (25 cases)	1	3	21
Lynn and van Heerden (1975) (29 cases)	0	3	26
Present series (26 cases)	1	4	21

Only 2 patients had had an episode of profuse diarrhoea with abdominal distension suggestive of enterocolitis. Both cases had responded to rectal washouts. One of these patients was among the 3 requiring anterior resection.

In a review of 501 cases of Hirschsprung's disease (Swenson *et al.*, 1973) collected over 26 years there was an incidence of 7·5% of cases that could be described as ultrashort segment. In the period under review in the present series 22 patients underwent Swenson rectosigmoidectomy for Hirschsprung's disease involving pelvic colon and longer segments. The high incidence of ultrashort segment can be attributed to an increased awareness of the condition and referral for investigation of patients who had been attending hospital for many years with constipation.

Anorectal manometry is a simple and in our experience a reliable method of investigation. In 2 patients where tracings were unsatisfactory but where the history was strongly suggestive of Hirschsprung's disease forceful anal dilatation was performed initially. This produced a good temporary response and was followed by myectomy.

Difficulties may arise in the histological interpretation of specimens when the segment involved is less than 3 cm long. The more extensive use of histochemical examination of these specimens may help to overcome this difficulty (Meier-Ruge *et al.*, 1972).

Anorectal myectomy is a simple procedure, and in this series free of complications, and can be combined with anterior resection when the length of segment is too long for myectomy alone.

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