Hirschsprung's Disease:

Report of a Case in a 39-Year-Old Adult

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CONGENITAL megacolon commonly is described as a disease of infants and children; nevertheless, on rare occasions, it is found in adults. In the past decade there have been reports of true Hirschsprung's disease in two adults over the age of 50 years. The basic pathologic concept of congenital megacolon has been adequately described and is well known. The following case report of Hirschsprung's disease, occurring in a healthy-appearing 39-year-old man, is presented.

Report of a Case

A 39-year-old white man was admitted to the hospital on October 17, 1960, with chief complaints of intermittent abdominal distention and chronic constipation, relieved by the periodic use of laxatives and enemas. The past history revealed that the patient had been constipated since birth. His condition was diagnosed as Hirschsprung's disease at one year of age. Since then he had relied entirely on mechanical means to empty the colon. In the first ten years of life, he had been troubled with excessive gaseousness and flatulence, associated with severe constipation. While serving in the armed forces in Europe, he spent three years in a German prison camp.

On physical examination, the patient proved to be a wiry, asthenic man, in no acute distress. The abdomen was asymmetrically distended, the distention being confined chiefly to the left side. This area was compressible and tympanitic. Harsh peristaltic rushes could be heard over the same region. Sigmoidoscopic examination revealed a sudden change in the caliber of the lumen of the rectosigmoidal area, where the bowel became capacious and atonic. Blood examination, urinalysis and excretory pyelo-

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showed that a significant number of ganglion cells were present. A temporary cecostomy was made for decompressive purposes.

The postoperative course was relatively uneventful. Approximately two months after his discharge the patient was feeling well and having two to four well-formed bowel movements a day. He is completely asymptomatic at the present time.

Comment

Varying degrees of severity are known to exist in Hirschsprung's disease. The severe form in infants carries a high mortality rate in untreated cases. Bartle reports an estimated mortality rate of 85 per cent in this group. A small percentage of patients, however, are able to go through life with periodic mechanical emptying of the bowel when the occasion demands. The degree and length of the involved bowel obviously determines the rapidity of progression of the disease.

The slowly progressive form might go undetected for years. On the other hand, the severe type requires early recognition and prompt surgical treatment to correct the abnormality. In both cases a rectal muscle biopsy, when properly performed, is the most reliable means of making a diagnosis. The frequent occurrence of aganglionosis involving the rectum and sigmoid makes this procedure feasible and practical.

Summary and Conclusions

True Hirschsprung's disease, occurring in the adult, is not reported frequently in medical literature. The patient presented

Fig. 2. Film made 24 hours later showing considerable retention of barium in the dilated sigmoid segment.

Fig. 3. Rectal muscle biopsy showing Auerbach's plexus with absent ganglion cells.