that he did not attain a full professorship at Harvard until he became emeritus in 1968.

REFERENCES


ABSTRACT OF ORIGINAL ARTICLE. The clinical syndrome known as Hirschsprung disease (HD), or congenital megacolon, had been recognized for more than a century. The pathogenesis of the dilation/hypertrophy of the colon without mechanical obstruction was puzzling, and the subject of great debate. Virtually all infants and children affected with this disorder died.

The authors studied 26 patients with congenital megacolon admitted to Boston Children’s Hospital during a 5-year period, from 1943 through 1947. Careful contrast radiographic study of the distal colon in all 26 revealed a functional obstruction produced by a dyskinnesia of the rectum. This was detected by running a small amount of barium slowly into the rectum with the patient in an oblique position. A portion of the rectum/rectosigmoid was consistently subnormal in caliber. Barium then was allowed to run through this narrowed area into the dilated colon. The authors interpreted the radiographic findings as evidence of a physiologic partial obstruction of the rectum/rectosigmoid.

Colostomies were performed in the dilated portion of the colon in 3 early patients, and all symptoms/signs of HD disappeared in each instance. These returned, however, with closure of the colostomies and persisted until the ostomy was reestablished. Colostomies then were performed in an additional 12 patients, and each was relieved of symptoms with restoration of a normal-caliber colon. The authors recognized that the distal obstructing segment would have to be removed before the colostomy could be closed.

They devised a special “pull-through” form of resection and tested it in the laboratory. The operation was performed subsequently on 23 patients, with no fatalities. Closure of the colostomy was possible in all but 2. The authors outlined the operation in detail in the article. Three infants, the youngest being 2 months old, tolerated a one-stage operation. The authors indicated that all patients subsequently demonstrated the urge to defecate and had normal bowel control.

Patients with HD have a physiologic obstruction in the nondilated segment of colon. Early colostomy relieves the symptoms/signs of obstruction, and removal of the narrowed segment results in cure.

COMMENTARY


Comments by Joseph F. Fitzgerald, MD

This watershed article described in fine detail the evaluation and management of patients with congenital megacolon. The authors included additional early observations in their article. They observed that the onset of symptoms was in the first week of life in all patients where early history was known (25/26). This was similar to the 2 patients described by Harald Hirschsprung in 1888.1 This supported a congenital origin. They noted that there was a marked male predominance (22/26; 84%). Despite the fact that impacted stool was present in the colon on abdominal examination, digital examination of the rectum revealed an empty vault in 24 of 26 patients (96%). This also was an observation shared by Hirschsprung. The absence of ganglion cells was not mentioned in the article, even though this fact was known at the time.

Frederick Ruysch is actually credited with describing the first case of HD in the medical literature in 1691.2 He described a 5-year-old girl with an “enormous dilatation of the colon” at autopsy. Hirschsprung delivered his classic paper 2 centuries later. Hirschsprung, as stated above, did note a distal site of constriction in an 11-month-old child who came to autopsy. Despite this, his attention was directed to-
ward inadequate defecation attributable to the congenital dilation and hypertrophy of the colon. The first comprehensive study of the neural elements in the myenteric plexuses of the colons of patients with HD was conducted by Dalla Valle in 1920. He, however, did not correlate the histologic aganglionosis in the distal colon with the clinical picture of obstruction. Tiffin, Chandler, and Faber are credited with the recognition of the relationship between the aganglionosis and the obstruction. They stated in 1940 that the “localized absence of peripheral neurons of the parasympathetic system causes a functional partial obstruction in the same region with secondary dilatation and hypertrophy.” Swenson and Bill published their initial experience with the special pull-through form of resection, which described 23 patients who also are included in the present article, in a surgical journal 1 year before the publication of this article. Interestingly, they proposed in this 1948 article that a full-thickness rectal biopsy should be performed to make the definitive diagnosis of HD. This makes it even more surprising that aganglionosis is not mentioned in the landmark article in Pediatrics.

Swenson published a follow-up article in Pediatrics in 1951 reviewing his experience with 82 patients. In this study, he also described a crude manometric study performed on 5 patients. Three small rubber tubes with balloons attached were inserted through a transverse colostomy and positioned so that one balloon was at the splenic flexure, one in the descending colon, and one in the rectosigmoid. The balloons were inflated with 10 mL of air and was attached to recording manometers. Two patients with transverse colostomies resulting from a right colectomy for gangrenous intussusception served as control subjects. The kymographic tracings revealed a progression of peristaltic waves from the transverse colon to the anus in the 2 control patients, and normal peristalsis in the dilated and hypertrophied colon in the patients with congenital megacolon without progression of the waves into the rectosigmoid. The distal balloon recorded only slight contractions, a fraction of the amplitude of the normal peristaltic waves in the proximal colon.

Swenson pointed out that the diagnosis of congenital megacolon may be difficult to establish in newborn and young infants, because dilation of the colon was usually minimal and the abrupt change in caliber from the sigmoid to rectosigmoid was not present. He did point out that it was not unusual for a newborn with congenital megacolon to present with the clinical appearance of small bowel obstruction with vomiting, abdominal distention, and failure to pass meconium, a triad of clinical findings that have since been highly suggestive of HD. In addition, he recommended in this follow-up article that cystometrograms be performed in all patients with HD, pointing out that half of his patients with congenital megacolon had atonic bladders with increased capacity. It was suggested that this was support for the possibility of additional autonomic nervous system dysfunction.

Twelve of the 82 operated patients, all younger than 3 years of age, suffered an acute illness characterized by vomiting, severe diarrhea, and abdominal distention with fever. During these episodes, the buttocks became excoriated, and anal sphincter spasm developed. Severe, even fatal, illness followed unless help was given. The treatment was the insertion of a large rectal decompression tube with gentle washing of the colon with normal saline solution. This was performed daily for 4 to 5 days. [Oh how I remember seeing patients in my earlier years who had a previous successful Swenson procedure and developed severe abdominal symptomatology in conjunction with an acute infectious process. They would suffer with anal spasm and could develop a fatal enterocolitis unless colonic irrigation with decompression was performed.] Swenson indicated that this complication was not seen in older patients. I am not certain that everyone ultimately agreed with this opinion.

Swenson experienced a child who had a recurrence of obstructive symptoms, and at reoperation, an 8-cm aganglionic segment was found that had been missed at the initial procedure. To avoid a future, similar mishap, “rapid sections” (ie, frozen sections) were made during surgery to identify the extent of the proximal aganglionic bowel during the year before submission of this article. He reported that “eighty-two patients with congenital megacolon have been relieved of their symptoms by this new surgical technic which consists of resection of the rectum and rectosigmoid without disturbing anal continence.”

Swenson provided follow-up on 200 patients treated for HD over a 10-year period in 1957. By now, he realized that an unacceptable number of operated patients developed one or more bouts of severe, occasionally fatal, enterocolitis. This led Swenson to make a technical revision in the pull-through procedure. The anastomosis of the properly innervated bowel was placed 1.5 cm from the anus in the initial operation. He now proposed that a partial internal sphincterectomy be performed. He recommended that the aganglionic segment be resected 2 cm from the mucocutaneous junction anteriorly and 1 cm posteriorly.

The apparent morbidity of the Swenson pull-through operation led to the development of several other operative techniques. Some of the earlier “new techniques” such as those proposed by State and by Rehbein never attracted a significant number of disciples and subsequently fell into disuse. Duhamel, looking for an easier and safer procedure, described a variant of the Swenson operation that avoided extensive pelvic resection and excluded, rather than resected, the rectum. In this operation, normally innervated proximal colon is anastomosed with the posterior rectal wall in a side-to-side manner. Stricture or a persistent septum often resulted in recurrent fecal impaction. This led Duhamel and others to make revisions in the original operation. Duhamel reported no late deaths in 37 patients.

Soave proposed a substantially different surgical technique aimed at minimal damage to the pelvic structures. He removed the mucosa of the narrow rectal segment, leaving a seromuscular cylinder with
normal blood and nerve supply. The normally innervated colon was brought through the aganglionic muscular cuff, allowing a redundant 10-cm portion to protrude through the anus. Twenty-one days later, when spontaneous healing was accomplished, the stump was resected by cautery. The operation was complicated by abscess formation within the seromuscular cuff, necrosis of the protruding bowel, and retraction of the colon stump. Boley modified the Soave operation by performing direct anastomosis of the anal verge during the initial anorectal pull-through operation, thereby avoiding all of the complications relating to the protruding colon stump. Both operations required long-term, daily anocutonic dilatations.

Limited distal anorectal myomectomy, performed by cutting a strip 0.5 to 1 cm wide through the anal sphincter and posterior rectal muscle up to a length of 14 cm, has been advocated for treatment of short-ultrashort-segment HD. Limited distal anorectal myomectomy, performed by cutting a strip 0.5 to 1 cm wide through the anal sphincter and posterior rectal muscle up to a length of 14 cm, has been advocated for treatment of short-ultrashort-segment HD.18,19

Today, more Duhamel (Duhamel-like) and Soave (Soave-like) procedures are performed than the classic or modified Swenson procedure; yet, it is hard to surpass or even match the results that had been obtained by Swenson over many dedicated years. He was certainly a blessing to patients with congenital aganglionosis coli.

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COMMENTARY


Comments by Samuel Lawrence Katz, MD

ABSTRACT OF ORIGINAL ARTICLE. Infants 1 to 3 months of age were immunized with a vaccine combining diphtheria and tetanus toxoids with pertussis organisms. The combination product was adjuvanted with alum. Infants were injected at four-week intervals between 1–3 months of age and boosted at 7–9 months of age. Serum antibody determinations were done 3 months after completion of the initial three administrations; again at 7–9 months of age, 6 months after completion of the original course; and a booster injection was given and final determinations obtained on serum collected at eight to ten months of age. Utilizing a variety of antitoxin and agglutination techniques, more than 90% of infants had effective antitoxin levels against diphtheria and tetanus on all three laboratory determinations whereas only 53% at 3 months, 51% at 6 months and 80% at 10 months had titers judged protective vs pertussis. It was therefore demonstrated that infants at this young age could respond satisfactorily with humoral antibodies against both diphtheria and tetanus toxoids but less so against pertussis.

COMMENTARY

This article, written by clinical investigators from the departments of pediatrics and bacteriology at the University of Rochester School of Medicine contributed important information from
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The online version of this article, along with updated information and services, is located on the World Wide Web at:

http://pediatrics.aappublications.org/content/102/Supplement_1/205