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Bowel Problems in Adults After Surgical Treatment for Childhood Hirschsprung's Disease

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Introduction

Hirschsprung's disease is a rare illness that people are born with (congenital). It occurs annually in about 1 in 5,000 live births. In Hirschsprung's disease there is a lack of nerve cells (ganglion cells) in segments of the intestinal tract located in the colon and/or rectum. The most commonly involved segment is the lower part of the colon and the rectum (rectosigmoid colon), but the entire colon may be affected (Figure 1). Ganglion cells are responsible for the normal wave-like motion of the bowel (peristalsis), and when they are missing the stool stops and an obstruction occurs.

The treatment is surgery to remove the abnormal bowel segment and restore bowel continuity. Several different methods are used to remove the abnormal segment, "pull-through" the remaining normal bowel, and re-attach it to the anus. The most common are the Soave, Duhamel, and Swenson procedures. Following surgical treatment, most children have a good outcome, but some have persistent bowel problems such as constipation, soiling, fecal incontinence, and inflammation in the colon (enterocolitis). These symptoms can impact the quality of life, which also needs to be addressed.

Diagnosis is usually made during the first few months or years of life. When diagnosed at this age, the most common symptoms are an inability to evacuate stools, failure to thrive, or progressive abdominal distention. Occasionally, however, the diagnosis of Hirschsprung's disease is not made until teenage years or adulthood. Most patients diagnosed as adults report symptoms from an early age, but others do not experience symptoms until adulthood. Most frequently, adults report constipation as the most dominant symptom.

The long term outcome (10–30 years) of surgically treated Hirschsprung's disease has not been systematically reviewed. Most children appear to have a good outcome following surgery. Some fully recover, some continue to suffer with mild symptoms, and some others may present with new onset of bowel symptoms in adulthood. Hirschsprung's disease, once

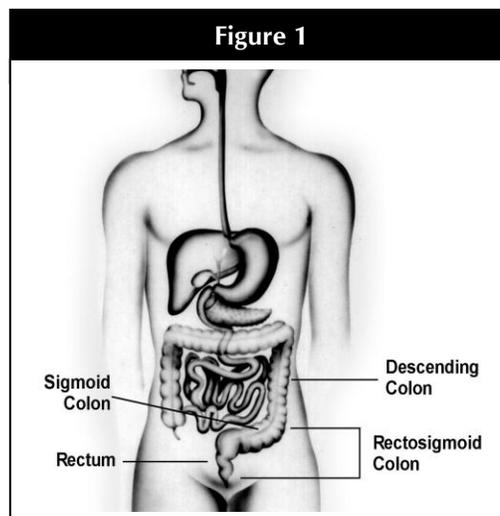
diagnosed, needs to be followed throughout the patient's lifetime.

Few doctors have experience treating adult patients with Hirschsprung's disease. As a result, the diagnostic and clinical approach to treatment is unclear. The following are descriptions of two patients seen in our clinical practice who experienced constipation and/or fecal seepage several years after their operation for childhood Hirschsprung's disease. We also describe the potential role of biofeedback therapy in the management of these patients.

Patient 1

History – Patient 1 is a 20-year-old woman. She was diagnosed with Hirschsprung's disease during infancy and underwent a Soave surgical procedure that consisted of removal of the sigmoid colon and rectum and attachment of the descending colon to the anorectum (Figure 1). Following this, her bowel habit had been fairly regular.

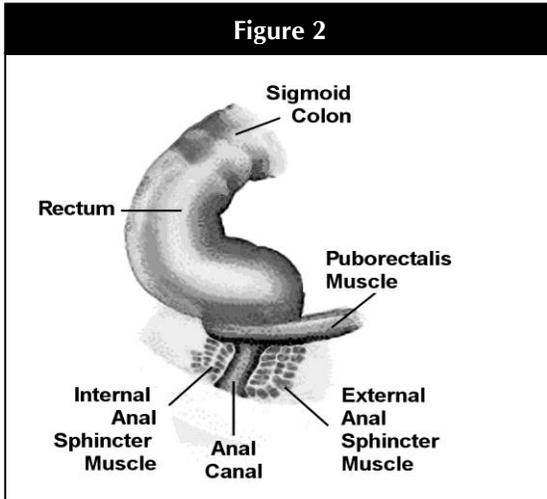
In her late teens, she began experiencing a change in her bowel habits. Previously, she had one bowel movement



every 2–3 days with a normal stool consistency. This changed gradually to bowels moving once every 7–10 days, and only after taking laxatives and with excessive straining. She visited the emergency room six times in one month, including two

admissions to relieve colonic distension. Abdominal x-rays and a CAT scan of the abdomen were normal apart from excessive stool.

Physical examination – Her general examination was for the most part normal. She did have increased resting and squeeze sphincter tone (tightness) with poor anal relaxation when asked to bear down. (Figure 2). The anal sphincter reflex contraction, in response to rectal distension, was normal.



At the lower part of the bowels, the sigmoid colon helps to slow the passage of stool before it moves into the rectum. The rectum can stretch to store material. It is surrounded by nerves that detect contents and signal a sensation of to let us know when it is time to have a bowel movement. When fullness is felt in the rectum, the puborectalis muscle and the external anal sphincter are voluntarily tightened to prevent leakage of stool. The internal anal sphincter is an involuntary muscle that helps to prevent leakage from the rectum. When it's time to have a bowel movement, all these muscles must relax so the stool can pass through the anus.

Investigations – Several tests of her bowel function revealed two abnormalities: 1) the time it takes for material to pass through her colon (colonic transit) was slow, and 2) she had *dyssynergic defecation*. Also termed *dyssynergia*, this is a condition marked by the failure of the pelvic floor muscles to relax with defecation. The pelvic floor is a group of muscles located at the lower part of the abdomen, between the hip bones, that supports pelvic organs, participates in defecation, and helps maintain continence. A *decrease* in muscle activity is necessary in order to have a normal bowel movement. Failure of this to happen can contribute to some forms of constipation, incomplete evacuation, and persistent straining with stool.

Patient 2

History – Patient 2 is a 25-year-old female who was diagnosed with Hirschsprung's disease as an infant and treated with a Soave operation at age two. During childhood, she had some difficulty controlling loose stools with occasional soiling.

At age fifteen, when she first visited our clinic, she had been experiencing stool seepage and intermittent lower abdominal pain for 2–3 years. Typically, she noticed stool staining of her underwear, but she was unaware of when the discharge occurred. Her bowels moved 2–3 times a day, sometimes with a normal urge to defecate and sometimes when she passed urine. She also had night-time soiling of her underwear. Her stool was normal in consistency. However, she could not differentiate between the passage of gas and stool.

Physical examination – Her general and abdominal examination was by and large normal. A digital rectal examination revealed decreased external anal sphincter tone (looseness) and a short anal canal. She was able to generate some voluntary squeeze pressure but when asked to bear down as if to defecate, she was unable to relax the anal sphincter muscles.

Investigations – Tests revealed that she too showed a dyssynergic pattern of defecation. She was unable to expel an artificial stool even after straining for five minutes. She had normal colonic transit and normal function in the main nerve going to the anal sphincter muscles (pudendal nerve). However, her rectal sensation (the way the body signals there is stool in the rectum and that it is time to go to the bathroom) was impaired. Leakage was observed during a test to measure rectal capacity (saline infusion test).

Treatment and Management Approaches

Both patients experienced persistent and unexplained symptoms and had made multiple visits to doctors or required hospitalization. In such patients, it is essential to understand the mechanisms causing their symptoms, and this is best achieved by performing appropriate physiological tests that access body functions and mechanics. With these tests we are able to evaluate and measure transit times in the colon, muscle pressure, rectal sensation, ability to retain and expel stool, and processes

of stool elimination. Using individually tailored programs, both dyssynergia and incontinence can be successfully treated.

Tests revealed that both patients had a lack of coordination of the abdominal, rectal, and anal sphincter muscles (dyssynergic defecation). In addition, Patient 1 had slow colonic transit; and Patient 2 had an impaired rectal sensation with an impaired capacity to retain saline. These findings enabled us to develop an appropriate treatment plan that consisted of lifestyle modification, advice from a registered dietician, and biofeedback therapy (neuromuscular training) that was tailored for each individual. Biofeedback is a painless neuromuscular reeducation tool in which a therapist, using special sensors to monitor functions that we are usually not aware of, helps to change abnormal functions to more normal and effective functions.

Patient 1 – The aim of biofeedback therapy for Patient 1 was to correct the lack of coordination or dyssynergia. She underwent three sessions of biofeedback therapy training. The training consisted of learning the anatomy and physiology of the pelvic floor, how to perform diaphragmatic breathing exercises, and how to appropriately relax the pelvic floor during defecation. She was asked to practice these techniques for at least fifteen minutes, 2–3 times a day at home, and to maintain a daily log of her bowel symptoms. The patient was also prescribed a medicine to relieve constipation. After the third training, she showed consistent relaxation of the pelvic floor enabling defecation. In addition, her bowel symptoms and stool consistency had improved. One year later, at follow-up, she reported complete satisfaction with bowel movements, a normal bowel habit, and only occasional use of laxatives.

Patient 2 – Patient 2 received advice on lifestyle modifications that included increasing fiber intake, paying attention to the feeling of the need to have a bowel movement (the call to stool), and attempting to pass stool at least twice a day after meals. Additionally, she received advice from a dietician.

She also received biofeedback therapy. The aim of the biofeedback was twofold: 1) to improve the dyssynergic defecation by promoting more complete evacuation, and 2) improve her ability to retain stools. Thus, she received instructions both for improving dyssynergia and for improving

continence by performing anal and pelvic muscle strengthening exercise. After five sessions of biofeedback training, she reported significant improvement in her bowel function. She had no night-time incontinence and her bowel movements became regular, at least 2–3 times a day. After the last biofeedback session, sensation and her capacity to hold and expel stool improved. At follow-up, one year later, her anorectal symptoms and bowel function had improved. Now, ten years later, she remains well, and has given birth to two normal children.

Conclusion

After definitive surgical treatment for Hirschsprung’s disease, some patients may develop symptoms during adulthood. It is important to follow-up Hirschsprung’s disease patients over a long term. A new onset of symptoms or persistent symptoms should be carefully evaluated and treated. Physiological and radiological (x-ray) tests can be of immense benefit.

Neuromuscular training with biofeedback, together with other behavioral or lifestyle approaches and medications, can be effective in the treatment of dyssynergic defecation or fecal incontinence in subjects with bowel symptoms after surgery for Hirschsprung’s disease.

Specialized Motility Tests

Gastrointestinal (GI) motility is defined by the movements of the digestive system, and the transit of the contents within it. When nerves or muscles in any portion of the digestive tract do not function with their normal strength and coordination, a person develops symptoms related to motility problems. GI motility tests measure different functions and provide doctors with important information for patient care. To learn more log on to IFFGD’s website at: www.aboutGI motility.org.

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