Defecation Disorders After Surgery for Hirschsprung's Disease

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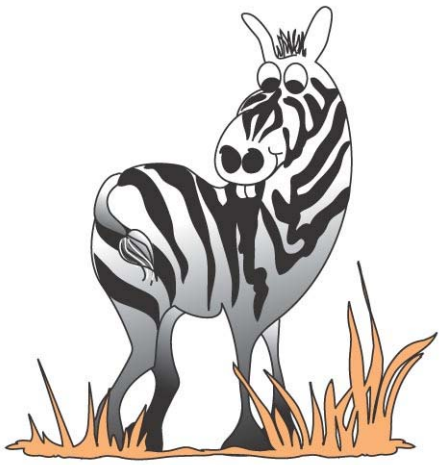

Over 1000 new cases of Hirschsprung's disease are diagnosed in the USA every year. More than half the children treated appropriately with surgery for Hirschsprung's disease suffer from chronic problems with constipation, incontinence, and/or abdominal pain. Even as adults, over half will experience occasional episodes of incontinence, and 10% will endure constipation unresponsive to medical management.

In the six decades since Ovar Swenson recognized that the distal bowel segment lacking ganglion cells was the diseased portion and created the first successful surgical technique, surgeons have been frustrated with the imperfect response to a surgery which eliminates the disease. Parents have been frustrated and children shamed by the inability to gain control over their bowel movements.

Over the past 10 years the pediatric motility community has discovered reasons and solutions for chronic post-operative problems. It is now clear that the child is not lazy or uncooperative, but has recognizable and treatable causes for the symptoms.

Constipation

There are three mechanisms for constipation: **functional constipation** (also called functional fecal retention), **neuropathy proximal to the aganglionic segment**, and **hypertensive anal sphincter**. It is not easy to distinguish among these conditions when they follow Hirschsprung's disease surgery. Only colon and anorectal manometry clarify the diagnosis and treatment.

Functional constipation

Functional constipation is the most common condition referred to pediatric gastroenterology clinics. It arises when an infant or toddler has a painful bowel movement. Infants and toddlers normally respond to a painful event by thereafter avoiding the event, like when they burn a finger on a hot stove. When they have a painful bowel movement, they attempt to avoid defecation, and after a few days have an even bigger, more painful bowel movement. This cycle of painful defecation and withholding behavior continues until the child experiences a sustained period of painless defecation.

Children with Hirschsprung's disease are even more likely to get functional constipation than healthy children, because they have so many different painful experiences around their bottoms. First, they get enemas and rectal studies before surgery. Next they have the surgery itself. Immediately following successful surgery, many children with Hirschsprung's disease have diarrhea for weeks. Diarrhea may break down sensitive skin under the diaper, and the child recognizes intense pain after each bowel movement. Because of the pain, infants learn to hold back their stools, and continue retentive behavior even after the diarrhea resolves because they fear the pain.

A few children need dilations when there is scar formation that narrows the new rectal lumen just above the anus. Dilations require that a surgeon or parent stretch the narrowing by running a few inches of a lubricated, smooth plastic or metal spindle through the anus. If dilations are done without proper analgesia and sedation, the painful, frightening event discourages the infant or toddler from defecation.

Sometimes surgeons suspect that a severe infection called enterocolitis is brewing inside, and so they order daily enemas and washouts to clean out the colon. These procedures, although possibly lifesaving, prevent the child from learning to control his bottom, and may provoke fear and consequently more retentive behavior. Then the infant's abdomen swells from retaining stool, and the surgeon worries more about enterocolitis, so that the cycle of fear (for clinician, parents, and infant) and pain (for the infant) is perpetuated.

The treatment for functional constipation is outlined in detail in another IFFGD pamphlet (Defecation Disorders in Children). Treatment begins with educating the child and parents about functional constipation:

- It is not dangerous.
- It is not a disease; it is a maladaptive but understandable response to painful stools.
- The colon cannot burst.
- Toxins do not leak back into the body from the colon.
- Non-stimulant laxatives are safe and effective.

Next, the clinician prescribes enough oral polyethelene glycol to melt away the mass of stool in the rectum over a few weeks. Polyethelene glycol is a crystal powder that dissolves odorless, colorless, and tasteless into the child's favorite drink, so that adherence to the prescribed dose is not a problem. It is not an emergency to get out the stool mass, and the more important goal is to return control of the child's bottom back to the child.

It is a good idea to see the doctor at least once a week for a few weeks, so that the doctor can listen to the parents' concerns and evaluate the stool mass by feeling the abdomen. There must be no rectal examinations, because rectal examinations frighten the child with functional constipation, and so interfere with treatment. Time is an ally. The child will let the stool out when he wants to do so. After the stool mass is gone polyethelene glycol is continued for many months, until the child and the parents are confident that acquisition of toilet skills is complete, and the fear is gone.

Neuropathy proximal to the aganglionic segment

Neuropathy proximal to the aganglionic segment is the second mechanism for chronic post-operative constipation. About 10% of all Hirschsprung's children have this problem. Although the aganglionic segment is resected and the remaining histology appears healthy, the nerve connections are not right.

Children with neuropathy proximal to the aganglionic segment have abnormal colon manometry. Sometimes there is a high pressure zone in one spot that does not relax to let colon contents flow by. Sometimes there are simultaneous contractions, but no coordinated, propulsive contractions. Sometimes there are no contractions at all. Most of the time the neuropathy is part of the Hirschsprung's disease, present from birth but unrecognized. Occasionally there may be a transient, treatable neuropathy due to the complications of inflammation after Hirschsprung's enterocolitis.

When there is a history of enterocolitis it may be worth a trial of anti-inflammatory drugs (eg sulfasalizine) or antibiotics (eg metrinidazole or clarithromycin). When there is no history of enterocolitis, one choice is using enough polyethelene glycol to keep stools watery for life, so that they flow without needing good motility. Another choice, better when the neuropathy involves a long segment of colon but the small bowel is spared, is surgery to remove all the diseased colon.

Colon manometry

Colon manometry discriminates functional constipation from colon neuropathy. Colon manometry is a simple test taking two or three hours in most cases, but preparation is complicated. First, the colon must be free of all solid stools. Cleaning out the colon may be unnecessary in a child with diarrhea, or a difficult problem in a child with a colon filled with hard stool. Cleaning out the colon may

be planned over weeks, so that it does not cause fear or pain. If it is necessary to clean out the colon quickly in an uncooperative child, it may be helpful to hospitalize the child for a nasogastric tube, a flexible plastic tube placed through the nose and throat and into the stomach, followed by medine given through the tube that causes bowel movements in rapid succession. In children who vomit easily a nasogastric tube may not work, and repeated enemas may be necessary.

Once the colon is cleaned out, the child is sedated. While he sleeps, the doctors place a thin plastic tube through his anus to measure the pressure in the anal sphincter. Then the doctors use a colonoscope, a plastic tube with a camera at the end, to look inside the colon as far as they can. Then they place a colon manometry catheter, a thin plastic tube with recording sites spaced 10 or 15 cm apart, through the anus to the beginning of the colon, and take out the colonoscope. They tape the catheter in place and confirm the catheter position with an X ray.

As the child wakes, the study begins. The child is encouraged to stay awake with peaceful, quiet-time activities. Parents are encouraged to comfort the child, and to ask questions about the study as they watch the results appear on the computer screen next to the bed. After an hour, the child eats lunch. Lunch normally causes an increase in colon contractions. In normal children and children with functional constipation the colon makes high amplitude propagating contractions (HAPCs), the physiologic marker for colon neuromuscular health. HAPCs are strong coordinated contractions that start at the beginning of the colon and move colon contents towards the anus before ending in the sigmoid colon, just before the rectum, the last portion of the colon.

Once HAPCs begin, it is most important to observe the child’s behavior: many children show distress, or stiffen and straighten their legs to hold it in. Sometimes you can point out to the child that the HAPC on the computer screen correlates with the urge to have a bowel movement or the pain that they feel. You can also tell them that what they are feeling is normal, and suggest that they allow themselves to have a bowel movement instead of holding it. In neuropathy, HAPCs and the increase in contractions after a meal are absent.

If there are no HAPCs before or after the meal, the doctors give bisacodyl, a stimulant laxative, through the catheter directly into the colon. In healthy children with functional constipation bisacodyl stimulates HAPCs in 10 or 20 minutes. In neuropathy, there are no HAPCs after bisacodyl. When the study is completed the catheter is withdrawn painlessly through the anus. Kids say the worst part of the testing is taking the tape off the IV and the catheter.

If HAPCs occur during manometry, the risk of enterocolitis is low, and chances are that the problem is one of failed toilet learning. When HAPCs are present, the best course is to stop doing all procedures that frighten the child, stop all

procedures through the child’s anus, and assure painless defecation. Moreover, it is often necessary to get a mental health professional familiar with Hirschsprung’s dsease and childhood defecation disorders to treat the child and help the family to cope.

Often the child is too frightened to have a bowel movement for many days. With each passing day the parents get more anxious that something is wrong. The child catches the fear from the parents, and the fear keeps him from having a bowel movement. If the doctor recognizes the symptoms and signs of bowel obstruction, and fears disease complications, he orders a battery of tests and treatments.

In fact, there is an obstruction, but it is exactly at the anal sphincter. The child is afraid to relax the anal sphincter. Frequent communications, confidence and trust among the parents, mental health professional and physicians are absolutely necessary to break the cycle of fear and withholding behavior.

Hypertensive anal sphincter

Hypertensive anal sphincter is the least likely cause of constipation, affecting about 5% of children with constipation after Hirschsprung’s disease surgery. In children with hypertensive anal sphincter, the anal sphincter pressures are so high, stool cannot pass even when there is an HAPC. Hypertensive anal sphincter is diagnosed by anal manometry when the child is asleep. If the child is studied while awake, there might be a false positive result because the child tenses up with fear when a tube is inserted through the anus to measure the sphincter pressure.

Occasionally a hypertensive anal sphincter may respond to botulinum toxin (Botox) injection. However, Botox treatment often fails, immediately or within a few months after a transient response. Surgical anal sphincterotomy is the best treatment for hypertensive anal sphincter, but sphincterotomy (cutting through the sphincter) is a delicate procedure. If the surgeon cuts too little there is no effect; if the surgeon cuts too much there may be lifetime stool incontinence. Therefore, sphincterotomy should be considered only if a hypertensive anal sphincter is documented by manometry under anesthesia.

Fecal incontinence

Fecal incontinence has more than one mechanism in post-operative Hirschsprung’s disease: functional constipation (functional fecal retention) and high amplitude propagating contractions through the neorectum. The rectum is a storage area for stools, and in Hirschsprung’s disease the diseased rectum has been removed.

In the healthy colon, stool moves several times each day when there is a very strong contraction that travels from the beginning of the colon all the way to the sigmoid colon, the area just above the rectum. When you have a healthy rectum these high amplitude propagating contractions dump the colon contents into the rectum. You get to decide whether to have a bowel movement or not, because your rectum can relax and stretch out to the volume.

After Hirschsprung's surgery, the rectum is gone, and HAPCs move colon contents to the anal verge with pressures that exceed the anal sphincter pressure. The child has two choices: relax the sphincter and experience incontinence, or hold tight as long as possible and experience intense crampy pain, the kind that you get when you have to go urgently but cannot get to the restroom.

There is another abnormality that makes incontinence even more likely in many children with HAPCs through the new rectum. When the colon is transected, there are more than the usual numbers of high amplitude propagating contractions. Instead of several HAPCs each day, there may be as many as several each hour. In health there are no HAPCs during sleep, but after colon transaction, HAPCs may occur during sleep, causing incontinence. Day and night incontinence improves in most instances with a small dose of amitriptyline at bedtime. Motility experts start with about 0.2 mg/kg/night, and increase by 0.2 mg/kg/wk until the problem stops or 1 mg/kg/night, whichever comes first. Amitriptyline reduces the pressure waves pushing out the stool, and reduces chronic pain.

The second cause of fecal incontinence is functional constipation (functional fecal retention). In functional constipation, a large stool acculmulates in the rectum. However, just like all of us, the child still needs to pass gas about 20 times every day. The sensitive anoderm can detect the difference between hard stool and gas, and between hard stool and liquid, but not between gas and liquid. Occasionally, some liquid stool oozes around the big, hard one. When the child relaxes the anal sphincter to pass gas, liquid that has seeped around the hard stool leaks out at the same time. Many school-age children will finally endorse this explanation for their incontinence, even though at first they try to avoid discussing or deny the problem. The treatment for functional constipation is discussed above.

Many children over 4 years of age blame absence of anal and colon sensation for incontinence. Sensation of the colon and anal sphincter are never affected by Hirschsprung's disease. The child may be having a problem with misunderstanding the signals coming from the bowels to the brain, but restoring the brain's recognition of normal sensations is part of the child's responsibility as he acquires normal toilet behaviors.

Abdominal pain

Abdominal pain may occur whenever the child senses an urge to defecate, but chooses to tighten the sphincter and avoid a bowel movement. Some children with a normal increase in colon contractions after a meal perceive those contractions as a pain rather than an urge to have a bowel movement. Over time the urge to defecate may change to a pain perception, so that the child no longer recognizes when it is time to have a bowel movement. This change in perception from a sensation that does not hurt to one that does is called hyperalgesia, and is caused by physiological changes in the pain nerves that run from the gut to the brain, and pain centers in the brain. A child who has had early life pain experiences, like surgery for Hirschsprung's disease, is at risk for developing hyperalgesia.

The abnormal nerve physiology is reversible, and there are effective treatments for visceral hyperalgesia. Drugs such as amitriptyline (in the same doses described above) and gabapentin slowly return pain nerve transmissions to normal in most children. For older children, cognitive behavioral therapy may be preferred to drugs in some families. CBT is a program that takes an hour each week, usually for about 10 weeks. CBT teaches the child to use the thinking part of the brain to take control of pain centers that may seem out of control. Some children benefit from a top down (CBT), bottoms up (drugs that reduce pain transmission to the brain) combination.

Outcomes

Parents worry that life for children with constipation or incontinence after Hirschsprung's disease surgery will not be as good as for other children. Parents worry that suffering from these problems, or years with a colostomy or ileostomy may worsen a child's personality, or quality of life later on. There is good news for these parents. Ajustment for teenagers and young adults with Hirschsprung's disease is no different than for healthy children. The number of interventions or amount of suffering did not correlate with getting along in life. The most important predictor of successful adjustment for children growing up with Hirschsprung's disease was unconditional love and acceptance by their families.

Currently only a handful of motility centers across the USA offer colon manometry. However, if your child with Hirschsprung's disease is not doing well after surgery, and if there are unanswered questions about diagnosis or treatment, IFFGD recommends that you contact a center of excellence in pediatric GI motility by phone or email, and get the answers to your questions before you make life-altering decisions.