

Managing post-surgery problems in children with Hirschsprung's Disease

By Marc A. Levitt, MD, Belinda Dickie, MD, PhD, and Alberto Peña, MD

Hirschsprung's disease is an inherited condition that causes newborn intestinal obstruction.

Children with the disease suffer dangerous intestinal blockages because they lack key nerves that drive the muscle contractions needed to move digested material. The disease occurs more often in males than in females, and sometimes is associated with Down syndrome and other inherited conditions.

Surgical treatments for Hirschsprung's disease have evolved over the decades. Surgery involves "pull-through" techniques to remove non-functioning portions of the bowel. Most surgeries are successful. However, there is very little published about the outcomes for patients who do not fare well after initial treatment.

Physicians at Cincinnati Children's Hospital Medical Center have studied more than 200 patients who had surgery at other institutions, then were referred to them with post-operative problems of fecal incontinence or abdominal distention and enterocolitis. Cincinnati Children's has developed a protocol for addressing these problems, which are fixable in almost every case.

Detailed evaluation is key

When a patient arrives with post-operative problems after a pull-through for Hirschsprung's, our protocol begins with a detailed history to determine bowel habits and to document reports of constipation, enterocolitis, abdominal distention, failure to thrive, incontinence, use of anti-motility agents, laxatives, or need for dilations or irrigations. Knowing the technique used during the original surgery is vital.

Patients also are examined under anesthesia to determine the integrity of the anal canal, assess for the presence of a stricture, assess the status of the sphincters, and look for the presence of a large rectal pouch or of a palpable Soave cuff. A full-thickness rectal biopsy also is performed.

These evaluations reveal two distinct subsets of Hirschsprung's patients with post-operative issues: 1) those who are soiling or fecally incontinent, 2) and those who have repeated episodes of abdominal distention and enterocolitis.

Incontinence can be managed

We define continence as the ability to have voluntary bowel movements without soiling and without the need for enemas. Ideally, fecal incontinence should not occur after pull-through surgery because patients are born with a normal continence mechanism, normal anal canal sensation and normal sphincters.

A key first step in managing incontinence is to determine whether patients' colons are hypomotile or hypermotile.

Hypomotility, slow-moving colon

Children with a dilated colon and a history of constipation are considered hypomotile. These patients are treated daily with a senna-based laxative. The amount of senna is adjusted over a period of days until the child has one to two soft bowel movements a day, with no accidents, and a radiograph showing a colon empty of stool.

Patients who continue to soil despite the laxatives are truly incontinent. These patients require daily bowel management with administration of large-volume (500 -750 mL), saline-based enemas.

Hypomotility, fast-moving colon

Children with a non-dilated colon are considered hypermotile. Rather than constipation, they have a tendency toward diarrhea. These patients are initially managed with loperamide, a medication to slow the colon; with pectin, a medication to firm the stool; and with a diet of constipating foods. Many patients achieve voluntary bowel control following this regimen.

Patients in this group who do not achieve voluntary bowel movements are incontinent. They require a daily, small-volume enema (200-500 mL) in addition to the colon-slowing therapy. These patients are re-evaluated every six to 12 months to determine if the enemas can be discontinued.

We have reviewed 130 patients referred to our institution's bowel management program following surgery for Hirschsprung's disease. Sixty-eight patients in this group had primary complaints of soiling.

Of those patients, 33 had dilated colons and a history of constipation. After initial treatment with laxatives, six patients became continent on laxatives alone. Of the 27 who did not respond to laxatives, 23 were managed with daily enemas.

Another 35 patients who were soiling had non-dilated colons and a tendency towards diarrhea. Six of these patients achieved voluntary bowel movements with medications and dietary changes. Of the remaining 29 patients, 23 were managed with a small-volume enema added to the therapy.

Five patients had damaged anal canals. They required long-term bowel management with enemas. Patients who have damaged or absent anal canals or damaged sphincters may also be good candidates for a Malone appendicostomy, a surgery to connect the appendix to the umbilicus and create a valve that allows enemas to be administered from the top of the colon instead of via the anus.

Re-do surgery for distention and enterocolitis

Abdominal distention following surgery for Hirschsprung's disease can be a sign of a dangerous condition called enterocolitis. In newborns, enterocolitis can be life-threatening and in its chronic form lead to failure to thrive.

In post-operative cases, enterocolitis can be due to pathologic or anatomic problems.

Assessment begins with a review of the patient's history, evaluating the anal canal under anesthesia and performing a rectal biopsy. We then implement a regimen of rectal irrigations with normal saline. Oral metronidazole often is added to the regimen if there is any evidence of enterocolitis, and sometimes must be administered for many months.

We then evaluate the underlying cause of the distention. Pathologically, two causes can result in abdominal distention and recurrent enterocolitis – pull-through of an aganglionic segment or pull-through of transition zone bowel. A rectal biopsy is performed to detect these problems.

Anatomic problems resulting in enterocolitis can be related to the initial operation. Three causes we have seen include stricture, a retained dilated segment of colon or a twist or kink in the pull-through segment. Each of these can be observed on a contrast enema.

All these problems can be corrected with a redo operation. Our approach is to perform a redo transanal resection with a Swenson technique to remove the problematic area. If the rectum is frozen with scar, a posterior sagittal approach may be needed.

We have performed re-operations on 75 patients who have had pull-through surgeries at other institutions. In all cases, the presenting symptoms were cured.

Conclusions

The ideal outcome for patients with Hirschsprung's disease is to achieve continence and successful stooling with the initial surgical treatment. However, when there is incontinence or distension, we can determine the cause through our evaluation. We then can take several steps to improve the outcome.

We can alter stool consistency and the colon's motility to enable some children to have voluntary bowel movements. Even when we cannot restore voluntary continence, we can have the child stop soiling in almost every case through bowel management and administering daily enemas.

And, in patients with persistent abdominal distention and enterocolitis, re-operations can significantly improve the quality of life.

Marc A. Levitt, MD, Belinda Dickie MD, PhD, Alberto Peña, MD, director of the Colorectal Center for Children at Cincinnati Children's Hospital Medical Center.

Division of Pediatric Surgery, Department of Surgery, University of Cincinnati.
3333 Burnet Avenue ML 2023
Cincinnati, Ohio 45229 USA

Telephone: 513-636-3240

Email: marc.levitt@cchmc.org