World Journal of Pediatric Surgery

Pullthrough pitfalls in treating Hirschsprung disease

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To cite: Rialon KL, Langer JC. Pullthrough pitfalls in treating Hirschsprung disease. *World Jnl Ped Surgery* 2024;**7**:e000862. doi:10.1136/wjps-2024-000862

Received 6 June 2024
Accepted 31 July 2024

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INTRODUCTION

Complications after a pullthrough for Hirschsprung disease can occur either early or late in the postoperative period. A small number of these patients may require reoperation, with rates varying from 3% to 28% at a large volume colorectal center.[1 2] Indications for reoperation include stricture, leak, fistula, transition zone pullthrough or retained aganglionosis, retained Soave cuff, or a Duhamel spur. Some of these may manifest as obstructive symptoms or constipation, which can be seen in 11% to 42% of patients.[3] Other patients may develop soiling and fecal incontinence, which can be caused by sphincter injury during surgery or by loss of the dentate line.[4] In a review of 46 redo pullthroughs, 71% occurred because of aganglionosis or a transition zone pullthrough, 19% from stricture or obstruction by the Duhamel pouch, and 8% from a tight Soave cuff.[2] The choice of pullthrough, whether a Swenson, Yancey-Soave, or Duhamel, does not influence the rate of complication but rather each has its own unique pitfalls to avoid.[5] The timing of surgery, whether performed as a neonate or delayed into infancy, continues to be debated, with a large study from the Pediatric Colorectal and Pelvic Learning Consortium (PCPLC) showing no difference in anastomotic or cuff stricture, Duhamel spur, or transition zone pullthrough, and several others showing higher rates of stricture and leak when repairs are done in the neonatal period.[6 7] In this review, we discuss avoidable pitfalls during the pullthrough procedure and provide guidance for identification and prevention.

EARLY COMPLICATIONS

Anastomotic leaks

Anastomotic leaks develop primarily from three possible causes: tension, inadequate blood supply, and technical error in performing the anastomosis. Leaks occur because of anastomotic dehiscence and patients may present with signs of a pelvic infection or sepsis, depending on whether or not they are diverted. Evaluation should begin with a contrast enema (figure 1) and exam under anesthesia (EUA) to assess the degree of leak/dehiscence. Cross-sectional imaging can be useful for percutaneous drain placement .

To avoid excessive tension on the anastomosis, the blood supply to the pullthrough segment should be carefully examined when taken to gain length. For transition zones in the sigmoid and more proximal, the inferior mesenteric artery or middle colic may need to be taken, which means the pullthrough will rely on blood flow through the marginal artery. The marginal artery is not always present along the entire length of colon, and one must take care to ensure it is present when taking the larger vessels.[8] The colon may need to undergo derotation for transition zones proximal to the mid transverse colon, as it may be difficult for the colon at this level to reach to the pelvis along the left side. Derotation also prevents obstruction of the duodenum by the pullthrough segment. This can be achieved by taking down all of the attachments from the transverse colon and greater omentum, followed by the hepatic flexure and a lateral to medial mobilization of the ascending colon. Once the colon down to the terminal ileum are free, the colon is brought down along the right side of the abdomen to the pelvis.

Traditionally, blood flow was assessed using Doppler probes and temporary clamping of vessels. Recently, the use of intraoperative indocyanine green fluorescence angiography (ICG-FA) has been found to be safe and feasible in cases of intestinal resection when needed to assess perfusion.[9] There are several fluorescence imaging systems available, with cameras for both open and laparoscopic procedures. The camera is positioned over the tissue to be assessed and ICG given intravenously at a dose range of 0.1–0.3 mg/kg. Within several minutes, the dye will reach the

Figure 1 Contrast enema demonstrating anastomotic leak (arrow).

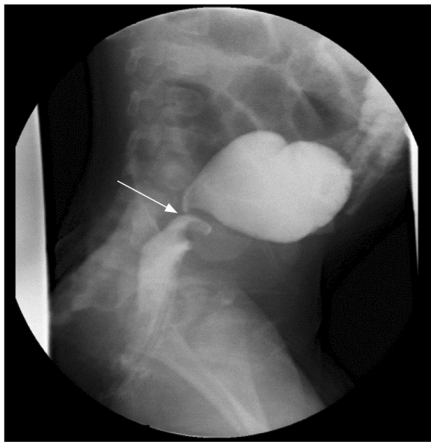

Figure 2 Contrast enema showing twisted pullthrough (arrow).

tissue and show viability of tissues in question. In a study of anorectal malformations and Hirschsprung disease, the use of ICG resulted in a change in the operative plan in 31% of cases.[10] The authors use this technology routinely to assess blood flow during more difficult pullthrough procedures, such as those with more proximal transition zones, those that require derotation, or reoperative cases.

A pullthrough procedure on bowel that has been chronically dilated can increase the risks of a leak due to the technical challenges associated with performing the anastomosis. This is of greatest concern in children diagnosed at a later age, as the bowel can be quite dilated and woody in consistency. The contrast study will provide information about bowel dilation. If the bowel is severely dilated, one should consider diversion to allow the bowel to return to a more normal caliber. Performing an anastomosis with dilated bowel results in a size mismatch between the proximal colon and relatively normal caliber anal canal. This can be addressed by using a "divide and conquer" approach, which consists of initially placing sutures in the 12, 3, 6, and 9 o'clock positions, and then dividing each quadrant sequentially to avoid gaps that might leak. Tapering of the proximal pullthrough can also be considered, although this creates an additional suture line that can leak. If the bowel is woody, it can also be more difficult to manipulate.

Recent bouts of enterocolitis can also compromise the anastomosis and increase risk of a leak. Patients showing any signs of enterocolitis should receive appropriate course of antibiotics and irrigations. The bowel should be allowed time to recover from the infection before attempting pullthrough procedure. Although there are no data to support a specific timeframe, the authors favor waiting until the completion of treatment for enterocolitis, when the child is back to baseline health.

Twisted pullthrough

Early obstruction postoperatively could be caused by a twist in the pullthrough segment (figure 2). This occurs when the proximal colon orientation is not maintained as it is pulled through the pelvis to the anal canal. This can vary in severity depending on the degree of misorientation. To avoid this when performing the pullthrough transanally, different colored sutures can be used to mark the antimesenteric and mesenteric borders of the colon every few centimeters.[11] A clamp can also be placed on the bowel and its orientation maintained throughout the dissection.[11] If performing the procedure with laparoscopic assistance, the colon can be visualized before and after it is pulled through to the perineum. Additionally, a chest tube or catheter can be inserted on completion of the anastomosis to make sure it can be advanced without obstruction.

LATE COMPLICATIONS

Stricture

Similar to anastomotic leaks, strictures can also be caused by poor blood supply and excessive tension. Some leaks, once healed, will develop into a stricture. Patients with a stricture may present with obstructive symptoms. Evaluation should begin with a contrast enema and EUA to assess the degree of stricture. Early identification of a stricture is critical, as early treatment may avoid redo pullthrough down the road. The surgeon should see the patient within a week or two after the pullthrough operation to calibrate the anus and consider routine dilations. Alternatively, the surgeon can calibrate weekly for 6 weeks.[12] If the child is unable to tolerate bedside dilations or there is concern for stricture on a digital exam performed in clinic, an EUA should be performed. Short, early strictures can often be managed with dilations under anesthesia with or without steroid injection or topical mitomycin.[11] Longer strictures or those refractory to dilation will require redo pullthrough.

Figure 3 Contrast enema with indentation of pullthrough segment (arrow) indicative of Yancey-Soave cuff.

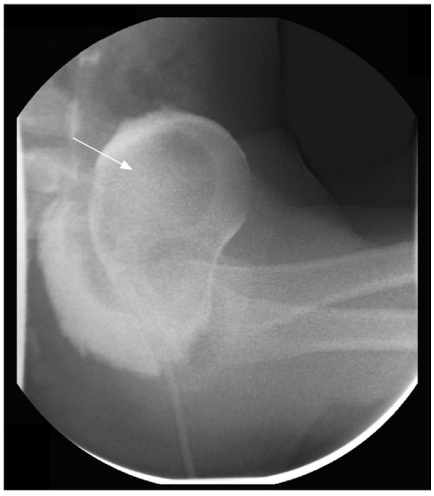

Figure 4 Contrast enema showing Duhamel spur (arrow).

Retained Yancey-Soave cuff

In the original description of the Yancey-Soave procedure,[13 14] the mucosal dissection was carried to the peritoneal reflection, which left a long cuff. This cuff can cause obstruction and its presence should be considered in the evaluation of any child with post-pullthrough obstructive symptoms.[4 15] A contrast enema will show a narrowing or indentation of the distal pullthrough (figure 3), and an EUA will demonstrate tightening extending beyond the anastomosis. Sometimes the extrinsic compression is proximal to the anastomosis, caused by the top of the cuff rolling down and causing a constricting ring. The best way to avoid this is by doing a short mucosal dissection of only 1–2 cm.[16] If creating a long cuff, it is important that it be divided or excised posteriorly. Care should also be taken to make sure the cuff does not roll down when bringing the pullthrough colon through the anal canal. The cuff size has been reduced over the last decade given the issues with longer cuff. Most now leave a cuff of 1–3 cm which does not need to be incised.[3 11]

As described for early stricture detection, those who underwent a Yancey-Soave procedure should have a digital exam in clinic to ensure the cuff is not obstructive. Routine dilations can be performed by parents or by the surgeon weekly. Yancey-Soave cuffs can sometimes be managed with dilatation alone, but in some cases, a redo pull-through is necessary, either using a transanal approach or a laparoscopic approach for more proximal obstructions.

Duhamel spur

To create the Duhamel anastomosis, the plane behind the rectum is developed, the posterior anal canal is opened above the dentate line, and the proximal colon is pulled through. An anastomosis is created between the ganglionic colon posteriorly and the aganglionic rectum anteriorly, usually using a stapling device inserted transanally. A Duhamel spur occurs when a portion of the aganglionic pouch above the anastomosis fills with stool and obstructs the proximal pullthrough.

To prevent this, several things should be avoided during creation of the Duhamel pouch. The aganglionated pouch should be kept short (the authors recommend no more than 4 cm). When creating the stapled anastomosis, multiple fires may be needed in order to reach the top of the pouch. At the completion of stapling, the common staple line should be palpated to ensure there is a relatively small amount of aganglionic bowel above it. In addition, a rectal examination should be done several weeks postoperatively, to make sure that the two staple lines have not coapted, which ultimately will result in fusion of the staple lines and a retained spur. If they have, the examining finger can be used to separate them.

A spur can be identified on either an EUA or contrast enema (figure 4), showing a dilated, blind-ending limb.[17] On EUA, the staple line should be palpated to make sure it has not coapted. A stapler can be used to divide this and open up the pouch.[18] If the pouch has dilated and is excessively long, it can be resected either transabdominally or transanally.

Retained aganglionosis and transition zone pullthrough

A retained aganglionic segment occurs when the pullthrough is either done in the transition zone or there was an error by the pathologist, which can occur either during the frozen section or at the time of final pathology.[19] The discrepancy between the frozen section and final pathologic diagnosis has been reported to be about 3%.[20] There can also be loss of ganglion cells after the pull-through, although this is a rare occurrence.[21] The surgeon should ensure their hospital has adequate pathologic support and that the on-call pathologist is comfortable reading frozen sections for Hirschsprung disease.[20] If there is inadequate support, one should consider avoiding frozen sections and send permanent sections instead. A proximal stoma can be performed while awaiting final pathology. Full thickness biopsies should be performed to confirm nerve cells in both submucosal and myenteric plexuses,

as there is sometimes a discrepancy in pathology between the two layers.[22]

Once ganglion cells have been confirmed, the final anastomosis should be proximal to the biopsy site to avoid placing it in a location without circumferential ganglion cells. While recommendations vary between 1–2 cm and 20 cm, the authors prefer to go approximately 5 cm above confirmed level of ganglion cells.[23] Because the distribution of ganglion cells is not consistent throughout the circumference of the specimen, the entire proximal end of the resection or ostomy should be sent and examined for any significant neuropathology.[24 25] Published guidelines recommend either examining a concentric donut or dividing the specimen into linear sectors.[25]

Sphincter injury and loss of the dentate line

Fecal incontinence postoperatively can be due to injury to the sphincter muscles during dissection in the anal canal, impairment of sensation from loss of the transitional epithelium at the dentate line, or a combination of the two. The primary mechanisms of sphincter injury include overstretching and going too low with the rectal dissection in a Swenson pullthrough, which will also result in damage to the dentate line. To begin the dissection in the anal canal, most surgeons will use either sutures at the mucocutaneous junction or the Lone Star retractor (CooperSurgical, Trumbull, Connecticut, USA) to visualize the dentate line. The advantage of the Lone Star retractor is that the pins can be advanced to just above the dentate line, which helps protect it from injury during dissection. Once the dentate line is visualized, a line should be drawn about 0.5–1 cm above it for newborns, and 1–2 cm above it in older children, sometimes referred to as the "purple line of Lee".[11 26] Sutures are then placed circumferentially along the line to use as traction and the dissection begun right below this. If one does not carefully protect the dentate line or begins the dissection too distally, both the sphincters and sensation from the transitional epithelium will be impaired.

Overstretching is a result of using excessive retraction, particularly with angled retractors, in order to visualize deeper into the anal canal. A nasal speculum can be used instead to gently visualize the dentate line.[11] The transanal pullthrough avoids entry into the abdomen, but some studies suggest that doing the dissection completely through the anal canal results in excessive stretching of the sphincter muscles in orders to gain adequate visualization of the dissection.[27] It is important to ensure that the dissection is done outside of the anus with downward traction rather than reaching up into the pelvis with anal retraction. If the transition zone is thought to be above the peritoneal reflection, or the child is older, and thus larger, it may be difficult to do the entire pullthrough transanally without going into the abdomen. A combined approach, using a laparoscopic mobilization down to the pelvic floor, will limit the amount of transanal dissection needed and avoid potential damage to the sphincter muscles.

Postoperatively, damage to the sphincter muscles can be noted on EUA, with identification of a patulous anal opening. EUA is also helpful to assess the quality of the dentate line. Awake anorectal manometry is a more accurate way to assess the sphincter pressure.[28] A technique for reconstructing the sphincter muscles has been described, with encouraging early improvement in bowel control.[29 30] This can be done after primary pullthrough or during redo pullthrough, where there is increased risk of sphincter injury and stretching.

CONCLUSION

Careful consideration to the technical pitfalls described above can avoid early and late postoperative complications after pullthrough surgery.

Contributors KLR contributed to study conception and design, data acquisition, analysis and data interpretation, drafting of the manuscript and critical revision. JL contributed to study conception and design, analysis and data interpretation, drafting of the manuscript and critical revision.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Not applicable.

Ethics approval Not applicable.

Provenance and peer review Not commissioned; externally peer reviewed.

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REFERENCES

1 Levitt MA, Martin CA, Olesevich M, *et al*. Hirschsprung disease and fecal incontinence: diagnostic and management strategies. *J Pediatr Surg* 2009;44:271–7; .
2 Ralls MW, Freeman JJ, Rabah R, *et al*. Redo pullthrough for Hirschsprung disease: a single surgical group's experience. *J Pediatr Surg* 2014;49:1394–9.
3 Langer JC. Persistent obstructive symptoms after surgery for Hirschsprung's disease: development of a diagnostic and therapeutic algorithm. *J Pediatr Surg* 2004;39:1458–62.
4 Saadai P, Trappey AF, Goldstein AM, *et al*. Guidelines for the management of postoperative soiling in children with Hirschsprung disease. *Pediatr Surg Int* 2019;35:829–34.
5 Nasr A, Haricharan RN, Gamarnik J, *et al*. Transanal pullthrough for Hirschsprung disease: matched case-control comparison of Soave and Swenson techniques. *J Pediatr Surg* 2014;49:774–6.
6 Kastenberg ZJ, Taylor MA, Durham MM, *et al*. Perioperative and long-term functional outcomes of neonatal versus delayed primary endorectal pull-through for children with Hirschsprung disease: a pediatric colorectal and pelvic learning consortium study. *J Pediatr Surg* 2021;56:1465–9.
7 Westfal ML, Okiemy O, Chung PHY, *et al*. Optimal timing for Soave primary pull-through in short-segment Hirschsprung disease: A meta-analysis. *J Pediatr Surg* 2022;57:719–25.
8 Mann MR, Kawzowicz M, Komosa AJ, *et al*. The marginal artery of Drummond revisited: A systematic review. *Transl Res Anat* 2021;24:100118.
9 Le-Nguyen A, Bourque CJ, Trudeau MO, *et al*. Indocyanine green fluorescence angiography in pediatric intestinal resections: A first prospective mixed methods clinical trial. *J Pediatr Surg* 2023;58:82–8.

10 Rentea RM, Halleran DR, Ahmad H, *et al*. Preliminary Use of Indocyanine Green Fluorescence Angiography and Value in Predicting the Vascular Supply of Tissues Needed to Perform Cloacal, Anorectal Malformation, and Hirschsprung Reconstructions. *Eur J Pediatr Surg* 2020;30:505–11.
11 Langer JC. Surgical approach to Hirschsprung disease. *Semin Pediatr Surg* 2022;31:151156.
12 Temple SJ, Shawyer A, Langer JC. Is daily dilatation by parents necessary after surgery for Hirschsprung disease and anorectal malformations? *J Pediatr Surg* 2012;47:209–12.
13 Soave F. HIRSCHSPRUNG'S DISEASE: A NEW SURGICAL TECHNIQUE. *Arch Dis Child* 1964;39:116–24.
14 Yancey AG, Cromartie JE, Ford JR, *et al*. A modification of the Swenson technique for congenital megacolon. *J Natl Med Assoc* 1952;44:356–63.
15 Dickie BH, Webb KM, Eradi B, *et al*. The problematic Soave cuff in Hirschsprung disease: manifestations and treatment. *J Pediatr Surg* 2014;49:77–80.
16 Nasr A, Langer JC. Evolution of the technique in the transanal pull-through for Hirschsprung's disease: effect on outcome. *J Pediatr Surg* 2007;42:36–9; .
17 Demehri FR, Dickie BH. Reoperative techniques and management in Hirschsprung disease: a narrative review. *Transl Gastroenterol Hepatol* 2021;6:42.
18 Langer JC. Repeat pull-through surgery for complicated Hirschsprung's disease: indications, techniques, and results. *J Pediatr Surg* 1999;34:1136–41.
19 Dasgupta R, Langer JC. Hirschsprung disease. *Curr Probl Surg* 2004;41:942–88.
20 Shayan K, Smith C, Langer JC. Reliability of intraoperative frozen sections in the management of Hirschsprung's disease. *J Pediatr Surg* 2004;39:1345–8.
21 Cohen MC, Moore SW, Neveling U, *et al*. Acquired aganglionosis following surgery for Hirschsprung's disease: a report of five cases during a 33-year experience with pull-through procedures. *Histopathology* 1993;22:163–8.
22 Kapur RP. Practical pathology and genetics of Hirschsprung's disease. *Semin Pediatr Surg* 2009;18:212–23.
23 Langer J, Caty M, de la Torre-Mondragon L, *et al*. IPEG colorectal panel. *J Laparoendosc Adv Surg Tech A* 2007;17:77–100.
24 Smith C, Ambartsumyan L, Kapur RP. Surgery, Surgical Pathology, and Postoperative Management of Patients With Hirschsprung Disease. *Pediatr Dev Pathol* 2020;23:23–39.
25 Veras LV, Arnold M, Avansino JR, *et al*. Guidelines for synoptic reporting of surgery and pathology in Hirschsprung disease. *J Pediatr Surg* 2019;54:2017–23.
26 Levitt MA, *et al*. Swenson-like approach for Hirschsprung disease. *J Pediatr Surg* 2013;48:2289–95.
27 Stensrud KJ, Emblem R, Bjørnland K. Anal endosonography and bowel function in patients undergoing different types of endorectal pull-through procedures for Hirschsprung disease. *J Pediatr Surg* 2015;50:1341–6.
28 Trinh TH, Nguyen NM, Lam KT, *et al*. Anorectal manometry findings in relation with long-term functional outcomes of the patients operated on for Hirschsprung's disease compared to the reference-based population. *Pediatr Surg Int* 2023;39:131.
29 Bokova E, McKenna E, Krois W, *et al*. Reconstructing the anal sphincters to reverse iatrogenic overstretching following a pull-through for Hirschsprung disease. One-year outcomes. *J Pediatr Surg* 2023;58:484–9.
30 Krois W, Reck CA, Darbari A, *et al*. A technique to reconstruct the anal sphincters following iatrogenic stretching related to a pull-through for Hirschsprung disease. *J Pediatr Surg* 2021;56:1242–6.