Case Report

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Surgical Research

Duhamel Spur in an Adult Male with High Anorectal Malformation and Hirschsprung's Disease

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ABSTRACT

Background: *Although a Duhamel Spur is a well documented phenomena. Literature on its frequency and management is limited. Furthermore, the existence of concomitant Hirschsprung's Disease (HD) and anorectal malformations (ARM) is exceedingly rare.*

Case Summary: *A 29 year old male with a history of ARM, HD, and urinary stoma, presented with fatigue and body aches. CT revealed a blind-ending large pouch in the left lower quadrant. According to records from Pakistan, in the OR, the appendix, cecum, ascending colon, and transverse colon were missing as an infant. A small segment of gut ended blindly behind the bladder over the middle of the sacrum. Histology of the terminal portion of the colon revealed the absence of ganglion cells, suggesting HD. When the patient returned to the OR at one year old he likely received both a Posterior Sagittal Anorectoplasty for his ARM and a Duhamel procedure for his HD. During the operation his urethra was injured, repaired and a foley catheter was left in place for an unknown amount of time. At the age of 12 the patient was unable to evacuate his bladder and he was given a suprapubic vesicostomy.*

Management: *We performed a preoperative colonoscopy. Upon entering the anus there was a smaller lumen and a larger lumen. In the OR the patient was placed in a lithotomy position for cystoscopy and stent placement. Next, we performed a midline laparotomy. The mass was under such tension it virtually sprang forth from the abdomen when the fascia was opened. We then fired a stapler multiple times across the neck of the mass where it arose off of the rectum with the sigmoidoscope in place.*

Outcome: *Patient was able to tolerate a diet on POD 5. Urology planned for outpatient internalization of nephrostomy tubes. The patient was discharged in stable condition.*

Keywords

Duhamel Spur, Anorectal Malformation, Hirschsprung's Disease.

Case Summary

A 29 year old male with a history of anorectal malformation (ARM), Hirschsprung's Disease (HD), and urinary stoma, presented with fatigue and body aches. CT revealed a blind-ending large pouch in the left lower quadrant. Patient was diagnosed with a Duhamel Spur. Patient underwent colonoscopy to better define anatomy and then underwent Spur resection with the aid of ureteral stents and sigmoidoscope via midline laparotomy.

Clinical Questions/Learning Objectives

- Formulate a diagnostic workup for an adult patient with a history of pediatric anorectal surgery.
- Develop a treatment algorithm for an adult patient with obstructive symptoms and a history or PSARP/Duhamel procedures.

Background

Although a Duhamel Spur is a well documented phenomena. Literature on its frequency is non-existent and management is limited to case reports. In one case, a stapler was fired transanally to eliminate the spur in a one year old [1], in another case surgeons utilized preoperative colonoscopy and the spur was resected laparoscopically in a six year old [2] while in another study three patients required a new anastomosis after the spur was stapled off [3]. The age of the patient and the nature of obstruction will dictate the ideal operative management. Nevertheless, obtaining a colonoscopy, especially in an adult patient in whom the complication is unclear should be strongly considered. In the case of a spur which is too large to be stapled transanally, as was the case in our patient, the use of intraoperative colonoscopy was also vital in identifying the common channel and also the appropriate level to resect the spur without narrowing the common channel. Although this could be accomplished in some patients laparoscopically the size and chronicity of our patient's spur with its subsequent adhesions would have made a minimally invasive approach impossible.

In a systematic literature review exploring the existence of HD and ARM, 38 articles reported 90 cases of HD coexisting with ARM from 1952 to 2013 resulting in an incidence of 2 % of this association. Associated syndromes were reported in 23 patients: Currarino syndrome in 11, Down syndrome in 8, Cat eye syndrome in 3 and Pallister-Hall syndrome in one case. There was a median delay of 8 months for the diagnosis of HD from the initial diagnosis of ARM [4]. This delay in our patient was unknown. In another systematic review the association of ARM and HD was uncommon (2.0-3.4%) but higher than in the general pediatric population (HD incidence 1/5000 [5] and ARM incidence 1/5000) [6]. There was also a high incidence of coexistence of ARM and HD with severe associated syndromes [7]. Another review found that in cases of aganglionosis beyond the rectal pouch and fistula, surgical intervention is needed and it is important to not use the distal rectal pouch or parts of the fistula in the reconstruction [8]. While another case report noted the importance of a Barium enema before the second stage of ARM surgery, as an abnormal shape may indicate the presence of concomitant HD [9].

Presentation and Diagnosis

Our patient presented to an outside hospital emergency room after a short course of Levofloxacin prescribed did not resolve his fatigue and body aches. In the ED, in addition to generalized body aches and fatigue he had leukocytosis (11.7), BUN/Cr of 31/3.17, urinalysis from stoma showed gross urinary tract infection, and GFR of 26 with unknown baseline. Patient denied emesis or constipation.

Patient received a CT scan of the abdomen which revealed a blind-ending, large pouch, in the left lower quadrant which was dilated, filled with debris, as well as severe bilateral hydronephrosis.

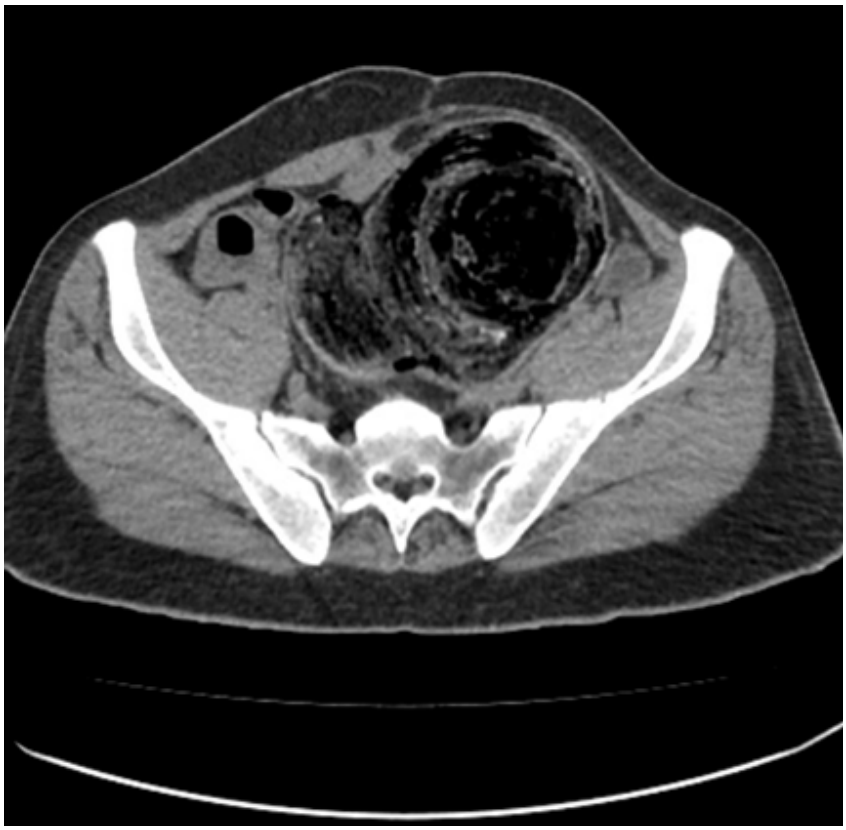


Figure 1: CT with blind ending, large pouch in the left lower quadrant.

Patient was started on IV Ceftriaxone and had bilateral nephrostomy tubes placed. Patient was discharged on PO Cefadroxil and instructed to follow up with a tertiary care center in order for more definitive management of the dilated loop of bowel in his left lower quadrant.

Patient presented to our colorectal surgery clinic several days after discharge from OSH where a more thorough surgical history was elucidated. Patient was born in Pakistan with both a high anorectal malformation and Hirschsprung's disease. On day of life two he was given a diverting loop colostomy. At one year old he returned to the operating room for loop reversal and definitive management of his high anorectal malformation. According to OSH records from Pakistan, in the operating room, the appendix, cecum, ascending colon, and transverse colon were missing. A small segment of gut ended blindly behind the bladder over the middle of the sacrum. Histology of the terminal portion of the colon revealed the absence of ganglion cells, suggestive of Hirschsprung's disease. It is unclear if this histology was taken at birth or at one year of life. We suspect that biopsies were taken during the loop colostomy procedure shortly after birth. When the patient returned to the OR at one year old he likely received both a Posterior Sagittal Anorectoplasty (PSARP) for his high anorectal malformation and a Duhamel procedure for his Hirschsprung's disease. During the operation his urethra was injured, repaired and a foley catheter was left in place for an unknown amount of time. 11 years later at the age of 12 the patient was unable to evacuate his bladder and he was given a suprapubic vesicostomy. At this time the patient and the family were offered further surgery for his hydronephrosis but the family refused any further intervention. When the patient presented to OSH at the age of 29 he reported that since the age of 12 he had voided both through his anatomic urethra and his suprapubic cystostomy. Only recently had he become obstructed and sick which prompted his trip to the emergency room.

After reviewing the imaging with our pediatric surgery colleagues we suspected that the patient had a Duhamel Spur. A Duhamel Spur is when a cuff of aganglionated bowel at the proximal end of the anastomosis is not incorporated into the common lumen. When this spur of bowel fills with stool it is unable to peristalse and slowly enlarges. In our patient it enlarged to the point that it caused bilateral obstructive hydronephrosis.

Timeline

- Day two of life: Loop colostomy
- One y/o: PSARP and Duhamel procedure
- 12 y/o: Suprapubic vesicostomy tube
- September 15, 2022 (29 years old): Bilateral nephrostomy tubes
- September 29, 2022: Colonoscopy
- October 6, 2022: Exploratory laparotomy, lysis of adhesions, resection of Duhamel spur

Management

We elected to perform a preoperative colonoscopy prior to operative intervention. Upon entering the anus there was a smaller lumen and a larger lumen.

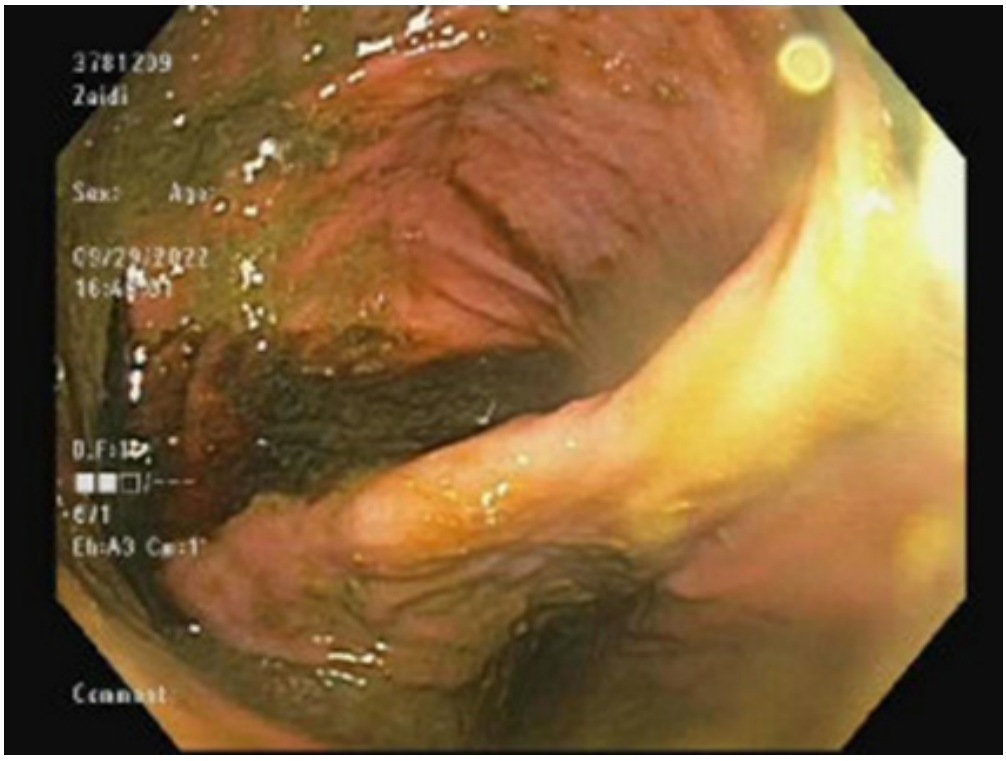


Figure 2: Dual lumen in colon with Duhamel Spur.

The smaller lumen was full of stool and the large lumen led to just 20 cm of colon prior to a normal appearing ileocecal valve.

After the colonoscopy we were confident that we would be able to resect the spur without stricturing the bowel and we scheduled the patient for elective cystoscopy, ureteral stent placement, exploratory laparotomy, and bowel resection the following week.

In the OR the patient was placed in a lithotomy position for cystoscopy and stent placement. Urology noted a stricture in the bulbar urethra which they were able to pass but were unable to identify the ureteral orifices during cystoscopy. They placed a 14 Fr. red rubber catheter in the suprapubic vesicostomy and a 16 Fr. Coude catheter in the urethra and left the nephrostomy tubes in place. Next, the abdomen was prepped and draped in standard fashion and a midline laparotomy incision was created. The mass was under such tension that it virtually sprang forth from the abdomen when the fascia was opened. The posterior aspect of it, however, was densely adhesed. We stayed close to the mass and dissected it circumferentially. The left-sided ureter was identified and kept out of harm's way. The right sided ureter seemed to be underneath the colon that had been brought down to do his anastomosis and we did not identify this for fear of injuring that portion of the colon.

After extensive dissection we performed flexible sigmoidoscopy to confirm that we would not be narrowing his original anastomosis. We then fired the Ethicon Echelon stapler multiple times across the neck of the mass where it arose off the rectum with the sigmoidoscope in place.

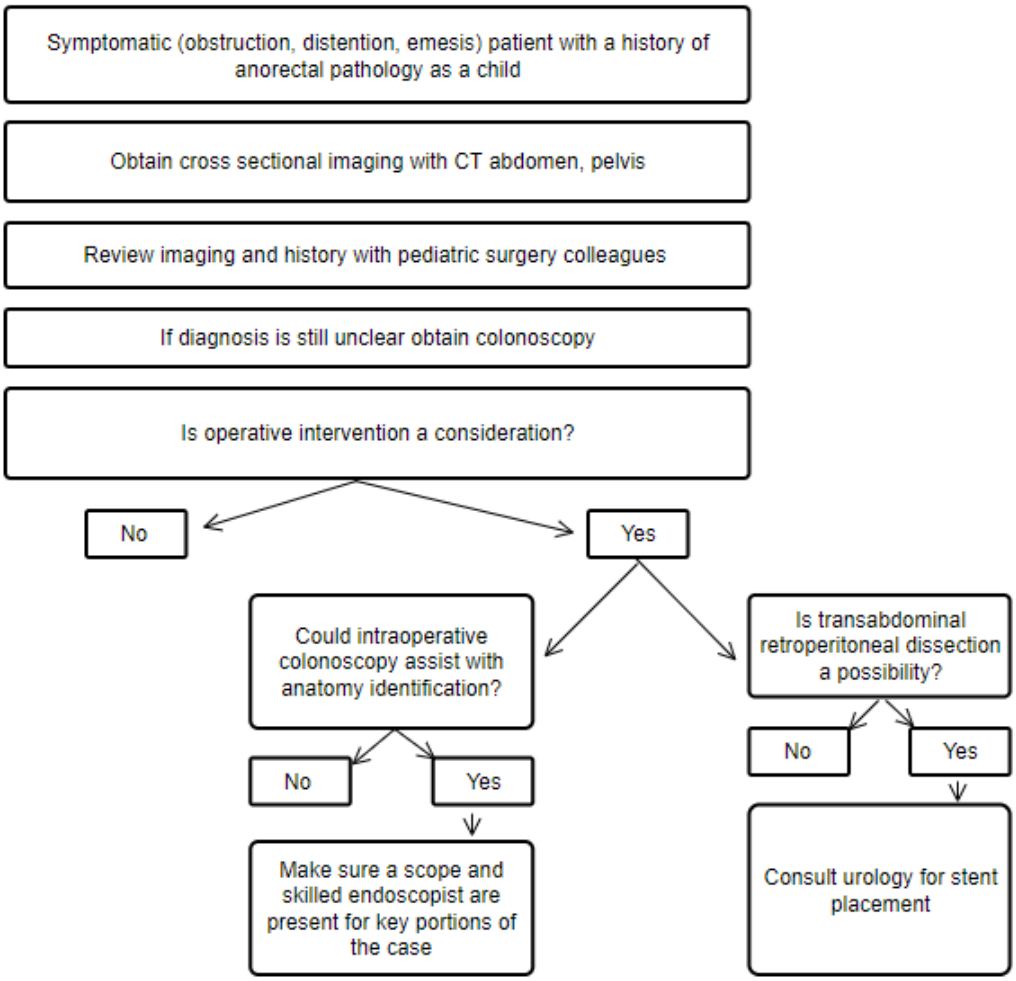
Next, the pelvis was instilled with saline and we then occluded the upstream part of the colon and performed flexible sigmoidoscopy again to be sure that all of the staple lines were watertight. We stayed close to the mass throughout the case, to avoid devascularization of any other structures.

19-Fr. Blake drain was placed in the pelvis and the small bowel was run. We then re-gowned and gloved and obtained a new set of instruments to close the fascia with 0 looped PDS. The skin was then partially closed to avoid a wound infection. After the skin was partially closed with gaps in between the staples, a wound VAC was placed in standard fashion.

Postoperatively the red rubber catheter was removed on POD 1, return of bowel function of POD 2, foley catheter was discontinued on POD 3 and he tolerated a diet on POD 5. Although he voided via his urethra he failed a nephrostomy tube (NT) cap trial on POD 5-6 and his NT's were placed back to drainage prior to discharge. Urology planned for outpatient internalization of bilateral NT's with interventional radiology and the patient was discharged in stable condition.

Figure 3: Spur is stapled off under direct intraluminal vision using a scope.

Evaluation and Treatment Algorithm



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