

Crohn Disease with Entero-Uracho-Cutaneous Fistula in a Pediatric Patient

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Background	A 16-year-old male patient with a known history of Crohn disease presented with an entero-urachocutaneous fistula.
Summary	A 16-year old male presenting with diarrhea, fatigue, weight loss, and abdominal pain was diagnosed with Crohn disease by pathology from a colonoscopy and cross-sectional imaging. He initially presented with abdominal pain, fatigue, and weight loss with feculent drainage from his umbilicus. Ultrasonography and computed tomography showed inflammation of the terminal ileum with a complex entero-urachal abscess, consistent with complex fistulizing Crohn disease. The patient underwent surgery to resect the terminal ileum, take down the fistula, and create a diverting loop ileostomy. Childhood-onset Crohn disease is associated with a wider extent of the disease and higher likelihood of requiring surgical intervention compared to adults. As such, pediatric Crohn disease patients pose a challenge to surgeons. The sparing of the noninflamed segment of the patent urachus as well as the bladder dome is unique to this case. The diagnosis and operative management of a rare manifestation of Crohn disease as well as the decision to opt for low-dose corticosteroids prior to surgery are highlighted by this case report.
Conclusion	Childhood-onset Crohn disease is associated with an increased prevalence of complications requiring surgical intervention. The authors present a pediatric patient with an entero-urachocutaneous fistula, highlighting the surgical management of this rare manifestation of Crohn disease and the role of low-dose corticosteroids in the pre-operative period.
Keywords	Crohn disease; enterocutaneous fistula

Disclosure Statement:

The authors have no conflicts of interest to disclose.

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Case Description

A 16-year old male presenting with diarrhea, fatigue, weight loss, and abdominal pain was diagnosed with Crohn disease by pathology from a colonoscopy and cross-sectional imaging. On his initial presentation he had severe terminal ileitis requiring hospitalization for treatment with high-dose steroids and an intraabdominal abscess requiring intravenous antibiotics, percutaneous drainage, and parenteral nutrition. Two months following his initial diagnosis of Crohn disease, the patient presented with feculent output from his umbilicus and from his previously placed percutaneous drain. Magnetic resonance enterography (MRE) confirmed the presence of a fistulous tract originating in his terminal ileum with communication to the skin of the umbilicus through a patent urachus. A follow-up contrast-enhanced computed tomography (CT) scan again demonstrated inflammatory changes in the terminal ileum with air tracking within the inflamed urachus (Figure 1). The patient was initially managed medically with antibiotics, prednisone, and infliximab. His symptoms continued to progress despite escalation of medical therapy, and thus operative management was performed.

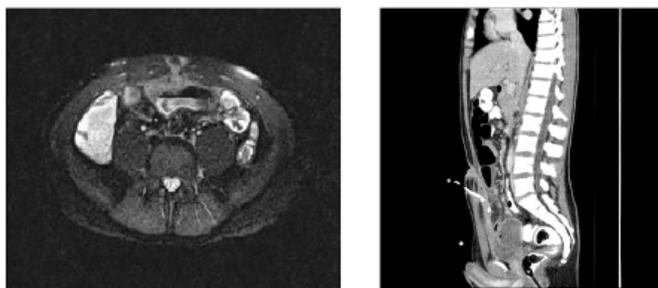


Figure 1. MRE and CT of abdomen/pelvis with contrast. Lobular 6.2 x 4.8 x 9.8 cm peripherally enhancing collection in the anterior pelvis extending from the dome of the bladder, extending to the anterior abdominal wall, and communicating centrally with the umbilicus.

A laparoscopic ileocolic resection with takedown of the entero-urachal-cutaneous fistula and diverting loop ileostomy was performed. Recognizing that the fistula entered the abdominal wall inferior to the umbilicus, a Hasson cut-down technique was used to place a 12 mm port superior to the umbilicus. A 5 cm segment of terminal ileum was the only area of inflamed bowel. The fistula was identified and dissected off the anterior abdominal wall with an energy device. The involved portion of the urachus was included with the specimen. The proximal and distal portions were closed with an energy device and chromic Endoloop. A 4 cm extraction site was created by extending the Hasson trocar site inferiorly. The bowel was exteriorized through a

wound protector. The bowel was sharply transected proximal to the diseased terminal ileum at an area of healthy bowel with soft, pliable mesentery and distally at an area of healthy ascending colon using the “pinch test” (Figure 2). The mesentery was divided between overlapping Kocher clamps and tied with interlocking suture-ligatures of #1 chromic sutures (Figure 3). The anastomosis was an end-to-side ileocolic anastomosis with a circular stapler (Figure 4). The colotomy was closed with a thoracoabdominal stapler (Figure 4). All staple lines were imbricated with 3-0 absorbable sutures. A proximal diverting loop ileostomy was created because of local sepsis, preoperative steroid use, biologic therapy, and malnutrition. The patient was weaned off parenteral nutrition prior to discharge and was sent home on postoperative day five after an uncomplicated course. Surgical pathology of the resected segment was consistent with active Crohn disease. The patient had a normal contrast enema study and returned to the operating room three months after the index operation for an uncomplicated closure of the loop ileostomy. He has returned to school and is gaining height and weight since that time. He currently has no signs of active Crohn disease and all wounds have healed.

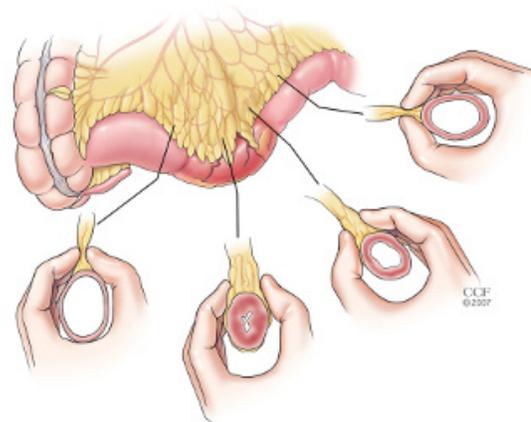


Figure 2. “Pinch” test to delineate diseased bowel from healthy bowel and mesentery. Reprinted with permission, Cleveland Clinic Center for Medical Art & Photography © 2007-2019. All Rights Reserved.

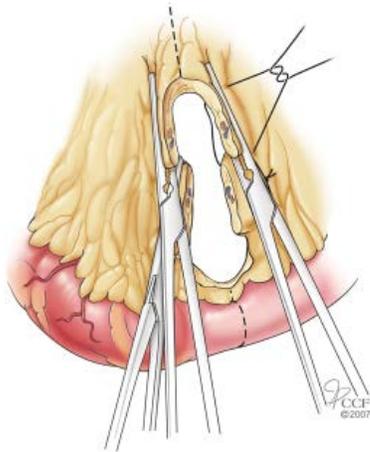


Figure 3. Division of the mesentery between overlapping Kocher clamps. Reprinted with permission, Cleveland Clinic Center for Medical Art & Photography © 2007-2019. All Rights Reserved.

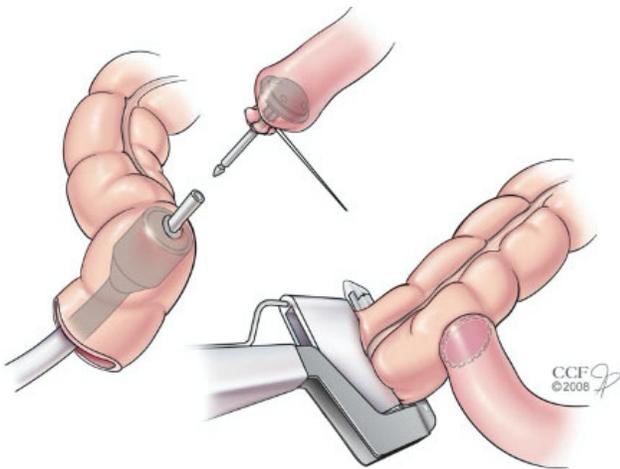


Figure 4. End-to-side ileocolic anastomosis with a circular stapler. Reprinted with permission, Cleveland Clinic Center for Medical Art & Photography © 2007-2019. All Rights Reserved.

Discussion

Childhood-onset Crohn disease (CD) is characterized by widespread extent, complications with early corticosteroid dependence, and need for surgical intervention in nearly half of patients by the age of 30 compared with 14 percent in adult-onset disease.¹ The incidence of CD in children and adolescents is increasing, with an incidence of 2.5 to 11.4 per 100,000 and a prevalence of 58 per 100,000.^{2,3} The characteristics of Crohn lesions along the gastrointestinal tract in children and adolescents is similar to those in adult-onset disease. In a large cohort of European children

with CD, ileocolonic disease was found in 53 percent of the patients, followed by isolated colonic disease (27 percent) and limited cecal disease (16 percent).⁴ The mainstay treatment, as with adult-onset CD, involves anti-inflammatory agents, biologics, and immunosuppressants with surgical interventions for complications including fistulas, strictures, abscesses, or medically-refractory disease.⁵ Careful selection of medical and surgical interventions are necessary given the high rates of complications and increased likelihood of repeat surgeries in the management of pediatric CD. Treatment priorities are given special consideration to pediatric patients compared to adults given their potential for growth and development.

This current case of CD presenting with an entero-urachal abscess from fistulizing disease of the terminal ileum with subsequent fistulization through the abdominal wall and patent urachus in a pediatric patient is unusual because the majority of Crohn fistulae are internal, most significantly entero-colonic, while external pathology (such as entero-cutaneous and entero-vaginal fistulae) are less common.⁶ Only three cases of entero-urachal fistula in pediatric CD patients have been reported to date.⁷⁻⁹ Previous reports advocated for en bloc resection of the fistulous segment and the bladder dome in diseases involving the bladder and manifesting with urinary symptoms.¹⁰ In this patient, the involvement of the urachus was limited to the segment near the umbilicus, sparing the bladder and the need for more extensive surgery, including complete resection of the urachus. Consideration was given to fully excising the urachus. However, on dividing the urachus, no open communication to the bladder was identified, suggesting that this segment was obliterated. Thus we did not feel the need to fully excise the urachus.”

This case also demonstrates the use of preoperative medical management of inflammation in a complicated fistula, especially the use of a low-dose corticosteroid regimen in the perioperative setting instead of the stress-dose regimen. Recent studies demonstrated minimal difference in outcome for inflammatory bowel disease patients receiving high-dose, low-dose, or no perioperative corticosteroids as well as a lack of advantage to stress dose corticosteroids.^{11,12} Thus, surgeons are shifting from the use of stress-dose corticosteroids, but practice patterns remain highly variable. In general, the use of steroids in children should be given special consideration due to their growth restricting effects, but they remain useful in the pre- and peri-operative setting in combination with antibiotics for decreasing complication risk.

Conclusion

Childhood-onset Crohn disease is associated with an increased prevalence of complications requiring surgical intervention. The authors highlighted the surgical management of a complex fistula presenting in pediatric Crohn disease and the beneficial role of low-dose corticosteroids in the preoperative period.

Lessons Learned

Childhood-onset Crohn disease is associated with an increased likelihood of requiring surgical intervention. The majority of Crohn fistulae are internal, most significantly entero-colonic, while external pathology (such as entero-cutaneous and entero-vaginal fistulae) are less common. Only three cases of entero-uracho-cutaneous fistula in pediatric CD patients have been reported to date. Surgical approaches in this unique setting may thus shift slightly from more standard techniques. In this setting, there is also a role for low-dose corticosteroids in the preoperative period instead of the use of a stress-dose regimen.

References

1. Ruemmele FM, Veres G, Kolho KL, et al. Consensus guidelines of ECCO/ESPGHAN on the medical management of pediatric Crohn disease. *J Crohns Colitis*. 2014;8(10):1179-1207. doi:10.1016/j.crohns.2014.04.005
2. Kappelman MD, Rifas-Shiman SL, Kleinman K, et al. The prevalence and geographic distribution of Crohn disease and ulcerative colitis in the United States. *Clin Gastroenterol Hepatol*. 2007;5(12):1424-1429. doi:10.1016/j.cgh.2007.07.012
3. Benchimol EI, Fortinsky KJ, Gozdyra P, Van den Heuvel M, Van Limbergen J, Griffiths AM. Epidemiology of pediatric inflammatory bowel disease: a systematic review of international trends. *Inflamm Bowel Dis*. 2011;17(1):423-439. doi:10.1002/ibd.21349
4. de Bie CI, Paerregaard A, Kolacek S, et al. Disease phenotype at diagnosis in pediatric Crohn disease: 5-year analyses of the EUOKIDS Registry. *Inflamm Bowel Dis*. 2013;19(2):378-385. doi:10.1002/ibd.23008
5. von Allmen D. Pediatric Crohn Disease. *Clin Colon Rectal Surg*. 2018;31(2):80-88. doi:10.1055/s-0037-1609022
6. Michelassi F, Stella M, Balestracci T, Giuliante F, Marogna P, Block GE. Incidence, diagnosis, and treatment of enteric and colorectal fistulae in patients with Crohn disease. *Ann Surg*. 1993;218(5):660-666. doi:10.1097/0000658-199321850-00012
7. Mador BD, Blair GK. Pediatric Crohn disease complicated by an entero-uracho-cutaneous fistula. *J Ped Surg Case Reports*. 2014;2:79-81. doi:10.1016/j.epsc.2014.01.006
8. Keir JA, McGregor R, Richards CJ, Windle R. An unusual presentation of Crohn disease. *Ann R Coll Surg Engl*. 2004;86(6):W22-W23. doi:10.1308/147870804137
9. Hollander LL, Girard ED, Ruscher KA, Sayej W, Kim C, Finck CM. Infected urachal cyst secondary to a Crohn enterourachal fistula. *J Pediatr Surg*. 2012;47(12):e43-e46. doi:10.1016/j.jpedsurg.2012.09.041
10. Tsukui H, Koinuma K, Morimoto M, et al. Crohn disease presenting as a ceco-urachal fistula. *Clin J Gastroenterol*. 2017;10(1):32-36. doi:10.1007/s12328-016-0691-2
11. Zaghiyan K, Melmed GY, Berel D, Ovsepyan G, Murrell Z, Fleshner P. A prospective, randomized, noninferiority trial of steroid dosing after major colorectal surgery. *Ann Surg*. 2014;259(1):32-37. doi:10.1097/SLA.0b013e318297adca
12. Zaghiyan K, Melmed G, Murrell Z, Fleshner P. Safety and feasibility of using low-dose perioperative intravenous steroids in inflammatory bowel disease patients undergoing major colorectal surgery: A pilot study. *Surgery*. 2012;152(2):158-163. doi:10.1016/j.surg.2012.02.019