

## Disorders of Anorectum in Pediatric Crohn's Disease: A Review Article

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### Abstract

Crohn's disease (CD) is an inflammatory bowel disease with chronic transmural inflammation of the intestinal wall and can be observed from oral cavity to anus. There is an increasing rate of CD in children. Surgery has a role in the management of this entity when medical treatment is unsuccessful. In this review article it is aimed to review the general characteristics of anorectal disease in children with CD and principles of management are discussed under the light of relevant literature.

**Keywords:** Crohn's Disease; Anorectal Lesion; Children

### Introduction

Crohn's disease (CD) is a progressive disease with extensive inflammation characterized by transmural involvement of tissues from mouth to anus. With an increasing incidence, particularly in children 10 to 19 years of age, it continues to be a challenge for both children with this disease and clinicians dealing with these patients. In this article it is aimed to review the general characteristics of anorectal disease in children with CD and principles of management are discussed under the light of relevant literature.

### Epidemiology

As an inflammatory bowel disease CD is commonly seen in children with an increasing incidence especially with age range of 10 to 19 years of age. It has been reported that 20% of patients with CD develop their inflammatory bowel disease before the age of 20 years [1]. Of these children, nearly one-third will have complications such as fistulae, strictures and abscesses within 5 years of diagnosis and compared to adults children with long standing disease have more risk of having surgery during the disease progression [2,3].

### Etiopathogenesis

The cause of pediatric Crohn's disease is not fully understood. There are multiple factors including genetic, immunologic and environmental parameters. It has been regarded as a result of malfunction of immunologic state of the host. Smoking and consumption of a more Westernized diet and frequent usage of antibiotics are associated with an increased risk of developing Crohn's disease. Responsible mutated genes in this entity include NOD2, ATG16L1, IL23R, and IRGM genes. It has been suggested that the genes in the interleukin 10 signaling pathway may cause Crohn's disease. Bacterial and viral infections are regarded as other environmental factors that cause Crohn's disease.

### Clinical features and definition of perianal Crohn's disease

Perianal disease in CD is a complex condition and is frequently refractory to treatment. It is present in 13 to 62% of children with CD [4,5]. These are in the form of fissures, skin tags, perianal abscesses and fistulae. At the time of diagnosis of CD 10% to 15% of children with CD have perianal fistulae [6-8].

Treatment options other than surgery include medications, nutritional and diet therapy. Corticosteroids and 5-aminosalicylic acid and antibiotics can be used. Immunomodulators such as 6-mercaptopurine, azathioprine and methotrexate may be used in the medical treatment of the disease. Drugs known as biologics including infliximab, adalimumab, vedolizumab and ustekinumab have been approved by the FDA for the treatment of Crohn's disease.

Abscesses should be treated promptly in order to avoid damage to the sphincter [9-11]. Most of the abscesses are small and difficult to drain and conservative treatment with metronidazole or ciprofloxacin can be enough for recovery. Drainage of greater abscesses can be performed via placement of a seton or large bore needle aspiration by ultrasonography or computerized tomography guidance or drain placement.

The treatment of the perianal fistula depends on the type of fistula. There are 2 types of perianal fistula in CD

1. Simple fistula: In this form there is superficial involvement of the external sphincter by single fistula tract without rectal inflammation or abscess.
2. Complex fistula: In this form there is deep or extensive involvement of the sphincter musculature by fistula tract or associated rectal inflammation, abscess or stenosis.

In the management of perianal fistulae it is recommended to locate fistula origin, depict the anatomical course of the fistula tract and to evaluate if there is rectal inflammation or abscess [7,9,12]. There are 5 consensus statements related to perianal disease in CD derived from guidelines from the paediatric inflammatory bowel disease Porto group of ESPGHAN [13]. These are:

1. Pelvic MRI and evaluation under anesthesia by a surgeon with experience in pediatric anorectal disease should be among the initial procedures in evaluating a child with suspected complex perianal CD (Agreement 100%)
2. Disease extension should be re-evaluated by ileocolonoscopy in all patients with complex perianal disease manifesting after primary diagnostic investigations of CD. Concomitant intestinal lesions (inflammation, stenosis) have both prognostic and therapeutic relevance. (Agreement 100%)
3. Treatment of perianal CD should be based on combination of surgery, antibiotics and biologics. (Agreement 100%)
4. Presence of perianal abscess should be ruled out early and when detected drainage should be discussed with the surgeon. (Agreement 100%)
5. Placement of noncutting seton should be considered in all complex fistula tracts, especially in those with recurrent abscesses. (Agreement 100%).

For the evaluation of perianal CD in children external and rectal examination followed by MRI imaging and examination under general anesthesia is sufficient [9]. Simple fistulae are located below the dentate line and have single opening and no associated stricture or abscesses. In the treatment of simple fistulae, together with antibiotic use including systemic and topical treatment, and surgical therapy

in the form of simple fistulotomy, or placement of noncutting seton, 80 - 100% of patients heal [13,14]. Uncomplicated fissures may be treated with topical therapy including sitz baths and topical ointments.

Complex fistulae involve superficial, transsphincteric or intersphincteric region below the dentate line have multiple openings and may be associated with rectal stricture or rectovaginal fistula [5]. In the management of these children aggressive immunomodulatory therapy in combination with surgical intervention is needed. During the management of complex perianal CD if there is concomitant abscess, it should be drained and antibiotics (metronidazole, ciprofloxacin) with immunomodulators such as anti-TNF $\alpha$  agents should be used in addition to surgical intervention. It is recommended that anti-TNF $\alpha$  agents should be continued for at least 1 year following with abscess drainage. It is also recommended that fistulectomy or fistulotomy does carry a risk of incontinence and it is suggested that these procedures be performed by an experienced surgeon [15]. A diverting proximal ostomy or proctectomy may be necessary for severe disease refractory to medical therapy. In children rectal stenosis or strictures are rather rarely seen and can be handled by repeated anal dilatation under general anesthesia.

There are newest approaches in the literature for the treatment of complex perianal disease such as use of fibrin glue and collagen to occlude fistulous tract with 75% resolution rate [16]. Endorectal flap advancement for complex perianal CD was also suggested to treat these cases with 46% resolution rate [17].

Prognosis in pediatric Crohn's disease depend upon several factors. It has been stated that patients with granulomas demonstrate a more severe disease course. Compared to simple fistulae, complex fistulae require more treatments and take longer to heal and have worse outcomes. Presence of a rectovaginal fistula is not thought to be a prognostic factor for overall perianal fistulae healing.

### Conclusion

Management of anorectal disease in children with CD may pose a great challenge for both the patients and clinicians dealing with these children. Although the number of children who require surgical intervention has decreased with the advances of medical treatment of CD, anorectal disease may produce anxiety for parents and children with CD. These children should be evaluated with external and rectal examination in addition to MRI and examination under anesthesia and when necessary a timely and appropriate intervention should be performed in order to avoid future complications and to gain future growth of the child with an acceptable quality of life.

### Conflicts of Interest

The author certifies that he has no affiliations with or involvement in any organization or entity with any financial interest, or non-financial interest in the subject matter or materials discussed in this manuscript.

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### Author Contribution to the Manuscript

Idea/concept, design, control and processing, analysis and/or interpretation, literature review, writing the article, critical review, references and materials by Volkan Sarper Erikci.

## Bibliography

1. Kappelman MD, *et al.* "Recent trends in the prevalence of Crohn's disease and ulcerative colitis in a commercially insured US population". *Digestive Diseases and Sciences* 58 (2020): 519.
2. Gupta N, *et al.* "Risk factors for initial surgery in pediatric patients with Crohn's disease". *Gastroenterology* 130 (2006): 1069-1077.
3. Vernier-Massouille G, *et al.* "Natural history of pediatric Crohn's disease: a population-based cohort study". *Gastroenterology* 135 (2008): 1106-1113.
4. Keljo DJ, *et al.* "Course and treatment of perianal disease in children newly diagnosed with Crohn's disease". *Inflammatory Bowel Diseases* 15 (2009): 383-387.
5. Surlin V, *et al.* "An update to surgical management of inflammatory bowel disease". *Intech Open* (2012): 1-28.
6. Griffiths AM. "Specificities of inflammatory bowel disease in childhood". *Best Practice and Research Clinical Gastroenterology* 18 (2004): 509-523.
7. DeZoeten EF, *et al.* "Diagnosis and treatment of perianal Crohn disease: NASPGHAN clinical report and consensus statement". *Journal of Pediatric Gastroenterology and Nutrition* 57 (2013): 401-412.
8. Muller KE, *et al.* "Incidence, Paris classification, and follow-up in a nationwide incident cohort of pediatric patients with inflammatory bowel disease". *Journal of Pediatric Gastroenterology and Nutrition* 57 (2013): 576-582.
9. Van AG, *et al.* "The second European evidence-based consensus on the diagnosis and management of Crohn's disease: special situations". *Journal of Crohn's and Colitis* 4 (2010): 63-101.
10. Haggett PJ, *et al.* "Pelvic and perineal complications of Crohn's disease: assessment using magnetic resonance imaging". *Gut* 36 (1995): 407-410.
11. Tang LY, *et al.* "Are perineal and luminal fistulas associated in Crohn's disease? A population-based study". *Clinical Gastroenterology and Hepatology* 4 (2006): 1130-1134.
12. Orlando A, *et al.* "The Italian Society of Gastroenterology (SIGE) and the Italian Group for the study of Inflammatory Bowel Disease (IG-IBD) Clinical Practice Guidelines: the use of tumor necrosis factor-alpha antagonist therapy in inflammatory bowel disease". *Digestive and Liver Disease* 43 (2011): 1-20.
13. Amil-Dias J, *et al.* "Surgical management of Crohn disease in children: guidelines from the paediatric IBD Porto group of ESPGHAN". *Journal of Pediatric Gastroenterology and Nutrition* 64 (2021): 818-835.
14. Nandivada P, *et al.* "Advances in the surgical management of inflammatory bowel disease". *Current Opinion in Gastroenterology* 28 (2012): 47-51.
15. Van Assche G, *et al.* "The second European evidence-based consensus on the diagnosis and management of Crohn's disease: special situations". *Journal of Crohn's and Colitis* 4 (2010): 63-101.
16. Lewis RT and Maron DJ. "Anorectal Crohn's disease". *Surgical Clinics of North America* 90 (2021): 83-97.
17. Regueiro M, *et al.* "Postoperative infliximab is not associated with an increase in adverse events in Crohn's disease". *Digestive Diseases and Sciences* 56 (2011): 3610-3615.

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