

Crohn's Disease: Clinical Presentation, Diagnosis, Treatment & Management

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Crohn's disease (CD) is a chronic idiopathic inflammatory bowel disease characterized by transmural inflammation and lesions that can affect the entire gastrointestinal tract, but most commonly occurs at the terminal ileum and colon. The prevalence of CD has an incidence of 3 to 20 cases per 100,000. The exact pathogenesis of CD is unknown, although a number of genetic, immune system and environmental factors have been shown to increase the risk of the illness and lead to the aberrant gut immune response characteristic of the disease. ¹

There is both direct and indirect evidence to suggest that genetics plays a role in the development of Crohn's disease. Direct evidence is provided as 32 different specific genetic alterations have been identified which are more common in people with Crohn's disease than in the population at large, and indirect evidence seen because Crohn's disease appears to run in families. ⁹ The immune system also becomes disrupted in Crohn's disease and sends tumour necrosis factor (TNF) antibodies to kill all bacteria, regardless of whether they are friendly or not. TNF antibodies cause most of the inflammation associated with Crohn's disease. ⁹

While Crohn's disease most often starts between the ages of 15 and 35, it can affect people of any age. CD affects men and women equally, and it appears to be more common in people in developed countries and among the Ashkenazi Jewish population. ¹⁰

Smoking is the most notable risk factor for developing Crohn's disease, aside from family history and ethnic background. Smokers are twice as likely to develop Crohn's compared with non-smokers and those who smoke usually experience more severe symptoms compared with those with the condition who do not smoke. While there is no evidence that diet causes, or plays a role in Crohn's disease, certain types of food and drink have been associated with worsening symptoms including milk, dairy products, alcohol, spicy, fatty and high-fibre foods. There is no single diet or eating plan that will work for everyone with Crohn's disease and dietary recommendations must be tailored individually. Although flares are sometimes associated with stressful events, there is no evidence to prove that stress causes or contributes to Crohn's disease. ⁹

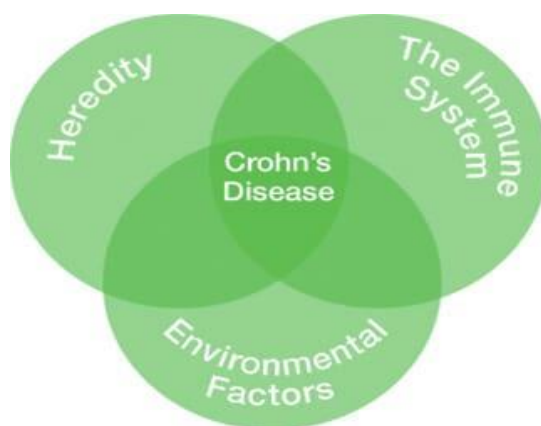


Image: <https://www.crohnsandcolitis.com/crohns/causes>

There are 3 main phenotypes of CD: inflammatory, structuring, and penetrating. Presenting symptoms are variable and some patients may have symptoms for years before the diagnosis of Crohn's disease is made. Patients with inflammatory disease often present with abdominal pain and diarrhoea, and may develop more systemic symptoms including weight loss, low-grade fevers, and fatigue. Patients with structuring disease may develop bowel obstructions while those with penetrating CD can develop fistula or abscesses. When an abscess is present, in addition to abdominal pain, patients can have systemic symptoms such as fever and chills and may also present with signs of acute peritonitis. ¹

Crohn's disease can affect any part of the gastrointestinal tract. Fifty percent of patients have involvement of the terminal ileum and colon, while 30% have small-bowel only involvement, and 20% of cases are isolated to the colon. In addition, 25% of patients suffer from perianal complications including fissures and fistula. Less frequently (<10%), patients present with isolated perianal complaints, upper gastrointestinal disease, or extra intestinal manifestations (EIMs) which can affect the skin, joints, eyes, liver, blood vessels, and kidneys. Arthritis is the most common EIM affecting up to 25% of patients with CD. ^{2,3}

Diagnosis

The diagnosis of Crohn's disease can be quite difficult given that presenting symptoms can be insidious and nonspecific. Symptoms that require further investigation include weight loss, bloody diarrhoea, iron deficiency, and night-time awakenings. Similarly, significant family history of IBD, unexplained elevations in the C - reactive protein level, sedimentation rates, or other acute phase reactants such as ferritin and platelets, or low B12 should prompt further investigation. There is no single test that can be used to confirm or disprove a diagnosis of Crohn's disease. The diagnosis of CD is made on the basis of symptoms, endoscopic and radiologic findings. (Colonoscopy, Biopsy, SBE, CT, MRI, Wireless Capsule Endoscopy) Pathology can also be confirmatory. ^{4,9}

Other conditions can mimic symptoms of CD so it is important to rule out infection and other causes even when patients with known CD are having flare-ups. Patients with diarrhoea should be assessed for infection, IBD, and in certain cases coeliac disease. Other conditions that may present similar to CD include appendicitis, Behcet disease, and Ulcerative Colitis. ⁴ Both **Crohn's Disease and Ulcerative Colitis** (UC) are inflammatory bowel diseases (IBDs), but there are some key differences. ¹⁰

Crohn's disease	Ulcerative Colitis
Inflammation may develop anywhere in the GI tract from the mouth to the anus	Limited to the large intestine (colon and rectum)
Most commonly occurs at the end of the small intestine	Occurs in the rectum and colon, involving a part or the entire colon
May appear in patches	Appears in a continuous pattern
May extend through entire thickness of bowel wall	Inflammation occurs in innermost lining of the intestine
About 67% of people in remission will have at least 1 relapse over the next 5 years	About 30% of people in remission will experience a relapse in the next year

It is important not to confuse inflammatory bowel disease (IBD) like Crohn's disease or ulcerative colitis with irritable bowel syndrome (IBS). IBS is a disorder that affects the muscle contractions of the bowel and is not characterized by intestinal inflammation, nor is it a chronic disease.¹⁰

Treatments

While there are several medications available to treat Crohn's disease, there is **no cure**.

Steroids

Steroids are used to induce remission but are not an effective maintenance agent. Steroids are usually only used to treat the active disease because their long-term use is associated with a range of adverse side effects. Budesonide and prednisolone are two steroids that are often used to treat Crohn's disease.^{1,9}

Budesonide is usually the first choice of steroid prescribed to help control the symptoms of Crohn's disease. Taken on a short-term basis of less than 12 weeks budesonide may cause acne, oedema of the face, hands, arms, feet and legs, mood changes, insomnia and indigestion. If taken for more than 12 weeks, budesonide may cause osteoporosis, increased vulnerability to infection, cataracts, muscle cramps and stiffness, and vitamin D and calcium supplements will be required to help protect against the effects of osteoporosis. Due to increased vulnerability to infections, close contact with people who are known to have infections, particularly those with chickenpox, measles and shingles should be avoided.⁹

Prednisolone is used in cases where budesonide proves ineffective. It has the same type of short and long-term side effects as budesonide, and has also been known to cause mental health problems in an estimated 5% of people.⁹

Aminosalicylates

Sulfasalazine belongs to a group of medicines called aminosalicylates, known to reduce inflammation inside the colon. Sulfasalazine can be used as an alternative to steroids to treat mild cases of Crohn's disease. Common side effects include headache, nausea, abdominal pain and diarrhoea.^{1,9}

Immunosuppressants

Immunosuppressants are used in maintenance therapy and in combination with steroids when a person has a relapse of symptoms. Two immunosuppressants widely used for treating Crohn's disease are azathioprine and mercaptopurine.

Common side effects of azathioprine include increased vulnerability to infection, bleeding and bruising. Less common side effects include headaches, shortness of breath, dizziness, nausea and vomiting.^{1,9,10}

Mercaptopurine originally developed to treat leukaemia has since proved effective in the treatment of Crohn's disease. Given in tablet form, its side effects are not as severe as those associated with other forms of chemotherapy treatment. Common side effects include nausea and vomiting while less common side effects include loss of appetite, fatigue, and

breathlessness and weakness caused by anaemia. Effective contraception if sexually active is important while taking mercaptopurine, as it temporarily affects both ova and sperm and can increase the risk of birth defects. ^{1, 9, 10}

Biological Therapies

Biological therapies are a new type of medication created using naturally occurring biological substances, such as antibodies and enzymes. Two main biological therapies are **infliximab and adalimumab**. Infliximab is usually only recommended in severe cases of Crohn's disease that have not responded to steroid and immunosuppressant treatments, and where the person is unsuitable for surgery.

Infliximab works by targeting the tumour necrosis factor (TNF) antibodies that are responsible for much of the inflammation associated with Crohn's disease. Given by intravenous infusion, around one in four people has an allergic reaction to infliximab and experiences symptoms such as joint and muscle pain, itchy skin, high temperature, rash, swelling of the hands and/or lips, problems swallowing and headaches. Symptoms range from mild to severe and usually develop in the first two hours after the infusion has finished. ^{1, 9, 10}

Adalimumab works in a similar way to infliximab by targeting TNF antibodies. It is given by injection and most people will need to have an injection every two weeks. Like infliximab, adalimumab increases vulnerability to infection, and those taking it should avoid contact with people with chickenpox or shingles and always report any symptoms of a possible infection to their GP. Adalimumab can cause reactivation of the hepatitis B virus and may not be suitable for people who were previously infected. Common side effects of adalimumab include pain, swelling, redness and itching at the site of the injection, headache, and abdominal pain, nausea, vomiting, skin rash, muscle, joint and bone pain and respiratory tract infections, such as colds and pneumonia. ^{1, 9, 10}

The goal of medication management is to control the inflammation and induce a clinical remission with pharmacological therapy, however, most patients will eventually require surgery. Unfortunately surgery is not curative and patients still require ongoing therapy even after surgery for disease recurrence. ¹

Surgery

Surgery is required when the symptoms of Crohn's disease cannot be controlled using medication alone. During surgery, the inflamed section of the digestive system is removed and the remaining part is reattached. An estimated 80% of people with Crohn's disease require surgery at some point in their life. Surgery does not cure Crohn's disease, but it can provide long periods of remission, often lasting several years. A stoma may be required if the disease or inflammation is so severe that an immediate anastomosis is not safe. In some instances, the stoma may be temporary and can be closed once the severe inflammation and infection is controlled. A colostomy is also required if the rectum is removed. ^{1, 9, 10}

Over 40,000 people in Ireland are living with inflammatory bowel disease (IBD), most of whom are diagnosed between the ages of 15 and 30. Of these approximately 20,000 people suffer from colitis and Crohn's Disease. Most Irish patients are hospitalised within two years of diagnosis and more than half will eventually need surgery. A report by the Irish Society for

Colitis and Crohn's Disease in 2015, showed that 49% of sufferers lost or had to leave a job because of their condition. The report also showed that Crohn's alone cost the health service €185 million a year and that paediatric cases of IBD have increased three-fold in Ireland since 2003. ^{5, 6, 7}

A survey of service providers treating patients with IBD, conducted by the Irish Society of Gastroenterology (ISG), found that the lack of dedicated specialist nurses in Ireland was the greatest barrier to delivering patient care. The Irish Society for Colitis and Crohn's Disease campaign for the 2018 World IBD Day (#DoubleUp) proved very successful. The campaign called for the number of specialist Inflammatory Bowel Disease (IBD) nurses to be increased from 14 to at least 28, to bring Ireland in line with international best practice. As of May 2019 the number of specialist nurses in Ireland has increased to 26. ^{7, 8}

A holistic person-centred approach is required in the management of patients with Crohn's Disease, who face a lifelong condition that can be emotionally and physically debilitating. The role of the clinician is to provide ongoing assessment, management, support and education. Key roles are to establish a therapeutic relationship with the patient, assess their understanding of the condition, establish goals and expectations for successful management of their condition and evaluate their physical, emotional, and psychological well-being. Common concerns for patients are adherence to their therapeutic regimen, pain and discomfort. Many patients experience anxiety, anger, frustration, stress and depression. Assessment, monitoring, audit and evaluation for disease activity, progression, and effects of the therapeutic regime on a patient with Crohn's disease is a continuous process. By implementing person-centred care, monitoring and evaluating symptoms, outcomes and responses to therapy, clinicians play a pivotal role in managing the illness and improving the patient's quality of life.

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