

Diagnosis and Management of Ulcerative Colitis

What is pediatric ulcerative colitis?

Ulcerative Colitis (UC) is an auto-inflammatory process that results in chronic inflammation of the large intestine. UC involves only the mucosa and extends proximally from the rectum. In children and adolescents, the area of involvement often extends beyond the splenic flexure. The extent of involvement does not correlate well with clinical disease activity. The mucosal depth and continuous distribution of the inflammation distinguishes UC from Crohn's disease in most cases.

Most researchers believe that UC is caused by a combination of genetic and environmental factors. Although UC may occur at any pediatric age, it is most often diagnosed in early adolescence. It affects males and females equally.

How does ulcerative colitis present?

The most common presentation of a patient with UC is bloody diarrhea and abdominal cramping. Extra-intestinal symptoms may also be present such as fever, skin rash, joint symptoms including frank arthritis, and liver disease.

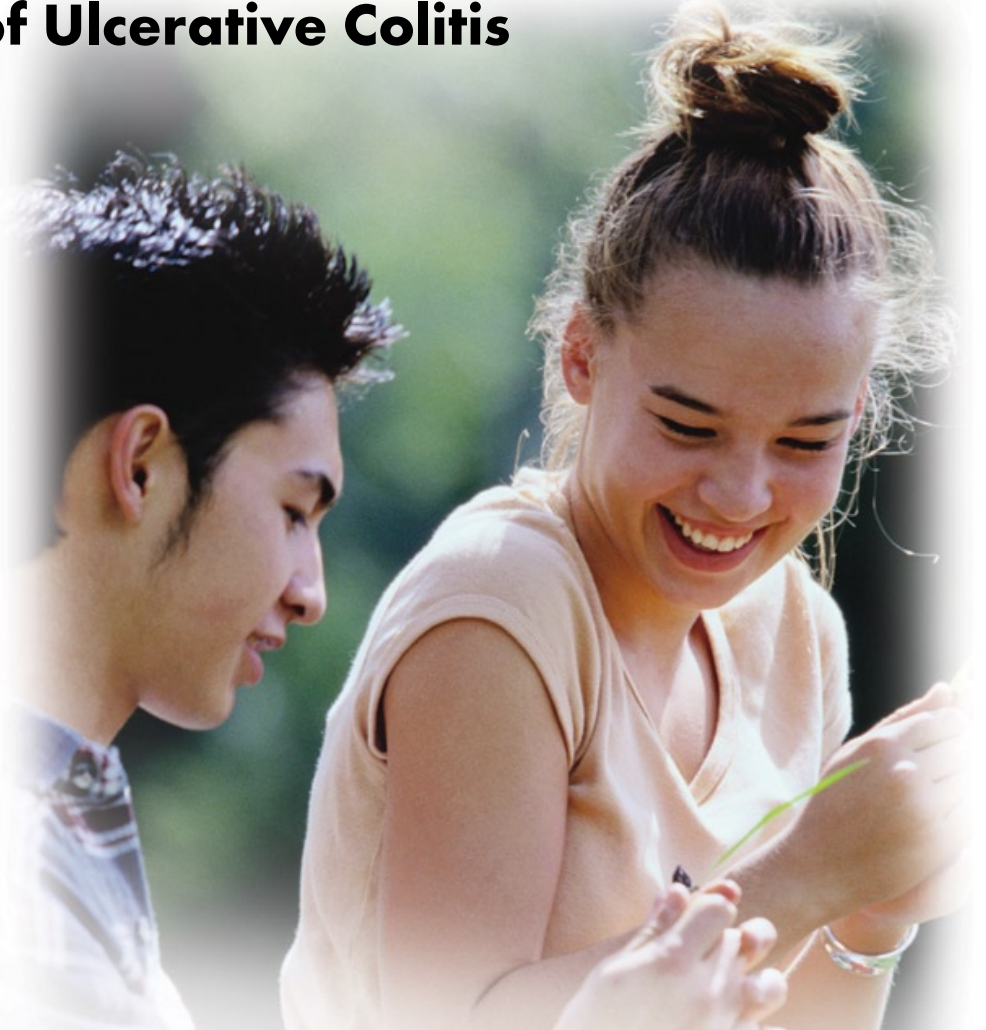
What evaluation should be offered by the primary health care provider?

Cultures for bacterial pathogens (Salmonella, Shigella, Campylobacter, Yersinia, E. Coli including O157:H7, Clostridium difficile toxins A & B) should be completed. Lab testing for parasites including serology for Entameba histolytica should be considered.

Additional testing (complete blood counts with differential, sedimentation rate, C-reactive protein and serum chemistries including albumin) should be part of the primary evaluation of the patient with persistent rectal bleeding. Anemia, elevation of acute phase reactants and hypoalbuminemia may be present indicating inflammation and chronicity. If the cultures are negative and the symptoms persist beyond two weeks, then referral to a pediatric gastroenterologist is appropriate.

When should I refer to a pediatric gastroenterologist?

Patients presenting with more insidious intermittent rectal bleeding, additional factors such as weight loss; extra-intestinal manifestations of inflammation and a family history of IBD should prompt early evaluation and referral.



In patients where the infectious evaluation is negative but bleeding persists with or without pain, referral is indicated.

How is the diagnosis made?

The diagnostic gold standard is a **colonoscopy** with biopsy. Grossly, a continuous colitis that begins in the rectum is the most common appearance of UC. Biopsies are taken to confirm the diagnosis and further exclude infections. It is not unusual for a pediatric gastroenterologist to also perform **esophagogastroduodenoscopy (EGD)** at the time of the colonoscopy since the EGD can help classify the diagnosis as one of Crohn's disease or UC. Additionally, a **contrast small bowel series** is frequently performed to image the rest of the bowel and to confirm that all the involvement is restricted to the colon and does not involve the small intestine, as is seen in Crohn's disease.

More recently, it has been noted that there are various antibodies that are seen in a majority of individuals with inflammatory bowel disease (IBD). While the diagnosis of IBD cannot be made serologically, antibody testing can help

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to confirm the diagnosis. Peri-nuclear anti-neutrophil cytoplasmic antibody (pANCA) is an auto-antibody that is most associated with UC and may help predict some potential long-term complications.

What treatments should I expect my patient to receive?

As with any chronic condition, the first goal of medical therapy in UC is to induce a clinical remission. Once remission is achieved, therapy is directed to maintain that remission.

Corticosteroids have been the mainstay of induction therapy for many years. The rapid onset of action makes steroids an attractive choice especially in patients who are ill and have a poor quality of life at the time of diagnosis. The immunosuppressive, cosmetic and metabolic side effects of steroids make them inappropriate to use as maintenance agents.

5-aminosalicylic acid (5-ASA) preparations are effective anti-inflammatory agents and have been demonstrated to both induce and maintain remission in UC. Clinical response can be somewhat slower than steroids and, therefore, patients with severe UC continue to receive steroids in most cases. Once remission is achieved, either with 5-ASA or steroids, maintenance therapy with 5-ASA is the most common therapeutic approach. There are oral and rectal preparations available.

Recurrent use of steroids becomes a concern in patients who are intolerant or unresponsive to 5-ASA. Since a goal of therapy is to eliminate long-term steroid use, **immunomodulator therapy** may be introduced. 6-mercaptopurine (6MP) and its parent compound Azathioprine are the immunomodulators most commonly used to steroid-spare in pediatric UC.

Most recently, biologic therapy in the form of infliximab (Remicade) has been approved for the treatment of refractory UC in adults.

In addition to medical therapy, psychosocial support of the pediatric patient and the affected family is critical in UC or any chronic pediatric illness. Implementation of a psychosocial support plan is a critical component of the health care plan. Often, the primary care provider has a unique and long term relationship with the family and child. This relationship can facilitate the execution of a coordinated psychosocial support plan

What are primary care issues in ulcerative colitis

• How important are vaccines?

It is important to recognize that children with ulcerative colitis continue to be at risk for routine childhood illnesses. Immunizations are an important protective mechanism and children and adults with ulcerative colitis should be maintained on the recommended immunization schedule.



At diagnosis, it is important that the immunization history be reviewed so that catch-up immunizations can be given if needed. Immunity to varicella should be confirmed by history or serology so that those who require varicella vaccine can receive the appropriate doses prior to receiving immunosuppressive therapy. The majority of vaccines do not contain live virus and, therefore, are generally safe even in individuals with ulcerative colitis who are on immunosuppressive therapy. All patients with ulcerative colitis should receive an annual vaccine for influenza. All patients with ulcerative colitis should be considered for an annual vaccine for influenza.

Live viral vaccines should be avoided in immune compromised children. This includes children on steroids (prednisone >20mg/d or 2mg/kg/day for 2 weeks or more), 6 MP/azathioprine, or methotrexate. Whenever possible, serologic conversion should be documented in children being immunized while immune compromised.

• When should I be concerned that the ulcerative colitis is flaring?

It is an old adage that patients “flare true”. The symptoms the child had at first presentation are generally the same symptoms at time of flare. Rectal bleeding would be the most common symptom of a UC flare. Other potential symptoms include diarrhea, fever, persistent abdominal pain or extra-intestinal symptoms including a rash, arthralgias/arthritis, jaundice or eye pain.

• When is it not a disease flare?

It is not always easy to distinguish between a viral illness and a flare of the ulcerative colitis. Time, patience and resolution of symptoms are often the determining factors. It is important to consider other sources for an illness prior to ascribing symptoms to the ulcerative colitis. Stool cultures and screening blood work looking for anemia, hypoalbuminemia or elevation of acute phase reactants can be helpful as well. Patients and their families can often tell the difference between an intercurrent illness and a flare of the ulcerative colitis.

First line anti-pyretic and analgesic therapy (for common fever, headaches and the like)

should probably be acetaminophen since there is some data to suggest that chronic NSAID therapy may adversely affect UC.

What is the role of surgery?

While medical therapy is the mainstay of treatment, surgery is performed in as many as 25% of pediatric UC patients. Steroid dependence and lack of response to medical therapy are the most common indications for colectomy in UC.

Advances in surgical techniques have made it possible to remove the colon and use the terminal ileum as a neo-rectum or pouch. The ultimate result allows for continuity of the gastrointestinal tract without a permanent stoma. An inflammation of the pouch can develop (pouchitis) and this can usually be treated medically.

How important is cancer surveillance in a UC patient?

It has been well established that patients with UC have an increased risk of colo-rectal cancer. Regular colonoscopy for cancer surveillance is recommended for all patients who have had UC more than 8-10 years including pediatric and adolescent patients. Surveillance of the pouch in patients who have undergone colectomy is usually recommended as well.

Physician Notes