Crohn Disease

Patient and family information, brought to you by the Education Committee of APSA

Overview - "What is it?"

Crohn disease is an inflammatory condition that can affect any part of the gastrointestinal tract from the mouth all the way to the anus. It typically affects all the layers of the intestinal wall. The most commonly affected area of the intestine is the last part of the small intestine (terminal ileum), but it can affect any part of the intestine and there will often be diseased areas along the intestinal tract with normal segments in between. The intestine can be affected in several ways. Sometimes, the intestine gets so inflamed that a hole (also known as a perforation) can result. This leads to a collection of pus (abscess) inside the belly. Sometimes, the intestine can become narrowed (stricture) or even blocked. Sometimes, different parts of the intestine get holes and create abnormal connections from one segment of the intestine to another (intestinal fistula) or an abnormal connection to the skin around the anus (anal fistula).

No one has really defined the real cause of Crohn disease. Experts believe that Crohn disease is because of many factors—inherited, environmental, and dietary, amongst others. The end result is that the body's immune system attacks the intestine causing the inflammation.

Crohn disease affects 0.7-15 per 100,000 people, with Caucasians being affected most commonly. A lower incidence is reported in Asia, the Middle East and is seen in the Southern hemisphere. Approximately one-third of people who develop Crohn disease are under the age of 20 years and the peak incidence is in patients 15 to 30 years old.

Signs and Symptoms - "What symptoms will my child have?"

The most common initial symptoms of Crohn disease include abdominal pain, diarrhea, decrease in appetite and weight loss. The location of the pain depends on which segment of the intestine is affected, so this can be different from one person to another. Sometimes Crohn disease can first present with perianal disease, including fissures (tears around the anus), abscesses (infections or pockets of pus) or abnormal holes drainage of with pus and stool.

Other associated symptoms are not related to the intestines at all. This supports the theory that the patient's immune system is attacking parts of the body without cause. Symptoms may include joint pain; tender, red skin nodules (erythema nodosum); red eyes with blurry vision; or inflammation of the liver. Children under the age of six are more likely to present with blood in the stool. As the disease progresses, children can have ongoing pain and diarrhea resulting in poor growth, low weight gain and delayed onset of puberty.

Diagnosis - "What tests are done to find out what my child has?"

Physical examination by an experienced healthcare professional is important.

Often times the diagnosis of Crohn disease is suspected based on the symptoms the child is having, the family history and physical exam findings. A full physical exam will include measuring height and weight, looking at the child's growth curve, a full abdominal exam, rectal exam and assessment of the eyes and skin. The surgeon may perform a more thorough rectal exam under anesthesia, especially if the child has complaints of pain, drainage in the anal area or problems with stooling.

Blood tests will look for evidence of infection or low blood count (anemia). However, there is no blood test that is currently available that confirms the diagnosis of Crohn disease.

Imaging can be helpful and may include CT scan, MRI, ultrasound and/or an intestinal contrast study (upper GI with small bowel follow through). What study is done depends on the symptoms. Your doctor will decide which one is best suited for the situation

Endoscopy is when a flexible telescope is placed in the mouth and is gently pushed through the esophagus, stomach, and part of the small intestine. The doctor can look at evidence of inflammation directly, but he will also obtain little pieces of tissue (biopsy) of the esophagus, stomach and first part of the small intestine. These pieces of tissue will also be sent for analysis under the microscope. Endoscopy is also done for the large intestine through the anal opening.

Conditions that mimic this condition: infectious enteritis (from bacterial infections), ulcerative colitis (another type of inflammatory bowel disease) or indeterminate colitis and appendicitis (inflammation of the appendix) can all have similar symptoms to Crohn disease.

Treatment - "What will be done to make my child better?"

Medical Options: Most of the time, medications are given primarily to decrease inflammation and decrease the symptoms. These medicines are used to target inflammation that is out of control. The medicines are NOT for Crohn disease only. In fact, you may have seen some of these medications used for other diseases that are caused by inflammation.

- **Steroids:** Steroids can be given by mouth, intravenously (through a vein), or as enemas. Some steroid names are hydrocortisone, budesonide or prednisone, amongst others. Steroids are very powerful anti-inflammatory drugs, but they are not preferred for long-term management due to side effects with prolonged use.
- **Non-steroidal** anti-inflammatory drugs: There are other anti-inflammatory medications that are not steroids. Some of these are mesalamine (Asacol, Pentasa), azathioprine (AZA), methotrexate and 6-mercaptopurine (6-MP).
- **Biologic agents:** Another class of medications are those that decrease inflammation by lowering substances in the blood that cause inflammation. These medicines are usually given as an infusion by vein every few weeks. Infliximab (Remicade) and adalimumab (Humira) are examples of these medicines.
- **Diet modification:** In some patients, a drastic change in diet is tried to see if this makes Crohn disease better. The child stops eating regular food and receives all necessary nutrition through a feeding tube with a liquid elemental formula (where the protein and sugars are broken down into the most basic components).

Many of these medications and diet changes are used in combination and at different time points throughout the disease process. The goal is to achieve control of the disease, decrease symptoms and promote intestinal healing.

Surgery: Surgery cannot cure Crohn disease. The role of surgery is to take care of complications of Crohn disease that cannot be helped with medication. When surgery occurs, the goal is to remove the least amount of intestine length. The usual management is to cut out the intestine that is narrowed or with a hole or abnormal communication and put the two ends back together. Sometimes, if the narrowing is short enough, the affected area can be widened up without removing the bowel. In some cases, especially those where there is infection, severe malnutrition, or high doses of anti-inflammatory medicines, a stoma may need to be formed. A stoma is a surgically created opening on the surface of the belly where a piece of intestine that is brought out to allow flow of stool. A bag is placed over this intestine to collect the stool. Depending on what needs to be done at surgery, the surgeon may decide whether a laparoscopic (several small incisions) or an open (one larger incision) approach is the best for your child.

Preoperative preparation: The most important part of preparing for surgery is making sure that the child has appropriate nutrition. The child will need to tolerate the surgery and heal appropriately. Nutrition is very important to decrease the complications of surgery. The patient may require additional oral (tube feeding) or intravenous nutrition both before and after the surgery, in some circumstances. Your child will be given fluids and antibiotics prior to surgery.

Postoperative care: The focus is on advancing the diet and administration of medications. Usually, the child is not allowed to eat after surgery because the intestines naturally slow down after an operation. He or she may require a tube through the nose into the stomach to drain it early after surgery. Once the child's intestines function, as evidenced by passing gas or stool,

then a diet will be started. Medications such as antibiotics, anti-nausea medication and pain medications will be given.

Short term risks include wound infection, abscess formation and small bowel obstruction. A wound infection happens less than 5% of the time (unless an infection is already present). Risk is increased if there is already an established infection or if the child is on high doses of anti-inflammatory drugs such as steroids. Infections may need only antibiotics or may require opening up of the wound. Abscesses are pus pockets inside the abdomen. If the abscess is small, antibiotics may treat it. If it is big, it may need to be drained. A small bowel obstruction results from internal scarring, which can cause the intestine to kink and become narrowed. The area where the intestines are sewn together can scar. Damage to surrounding organs can also occur, especially if the child has had several previous surgeries or the inflammation is severe. Damage can occur to other portions of intestine, the bladder, the fallopian tubes, or the uterus due to scar tissue that may have formed.

Long-term risks include recurrence and the development of cancer. Recurrence (the disease comes back) is very common in children undergoing bowel resection for Crohn disease. IT IS VERY IMPORTANT TO FOLLOW-UP WITH THEIR GASTROENTEROLOGY SPECIALIST TO RESUME THE MAINTENANCE MEDICATIONS TO KEEP THE INFLAMMATION UNDER CONTROL AND REDUCE THE CHANCE OF RECURRENCE. Children with Crohn disease are also at increased risk of developing a cancer of the colon or the small bowel later in life.

Benefit of surgery is fixing a problem that has been difficult to treat with medication. For example: a stricture leading to an obstruction, a fistula resulting in abnormal drainage, infection, or pain. Often when symptoms improve and disease has been controlled with surgery, the child will have an improved appetite, gain weight, and grow.

Home Care - "What do I need to do once my child goes home?"

Diet: After surgery the diet will be slowly advanced back to solids. There are options for dietary supplements that are thought to help with minimizing inflammation including omega-3 fatty acids (fish oils), short-chain fatty acids and other types of fiber. Your doctor may make suggestions as to which supplements are needed at home.

Activity: Your child should avoid strenuous activity and heavy lifting for the first 1-2 weeks after laparoscopic surgery, 4-6 weeks after open surgery.

Wound care: Surgical incisions should be kept clean and dry for a few days after surgery. Most of the time, the stitches used in children are absorbable and do not require removal. Your surgeon will give you specific guidance regarding wound care, including when your child can shower or bathe. If a child has an ostomy there will be teaching on how to care for the stoma prior to discharge from the hospital.

Medicines: Medicines for pain such as acetaminophen (Tylenol) or something stronger like a narcotic may be needed to help with pain for a few days after surgery. Stool softeners and laxatives are needed to help regular stooling after surgery, especially if narcotics are still needed for pain. Sometimes, if an ileostomy (portion of the small intestines is surgically connected to the abdominal wall) is needed, medicine to slow down the intestine and fiber is needed to decrease fluid loss. MEDICINES TO CONTROL CROHN DISEASE will be resumed by the gastroenterologist.

What to call the doctor for: Call your doctor for worsening belly pain, fever, vomiting, diarrhea, problems with urination, or if the wounds are red or draining fluid.

Follow-up care: Your child should follow up with his or her surgeon 2-3 weeks after surgery to ensure proper postoperative healing. Follow up with the gastroenterologist to discuss long-term management and recurrence prevention.

Long Term Outcomes - "Are there future conditions to worry about?"

Crohn disease is a life-long disease, and patients will have periods of remission (no symptoms) alternating with flare-ups of symptoms. It is possible that further surgery will be required if complications from the disease develop. It is important to establish long-term follow-up care with the child's pediatrician and gastroenterologist so that symptoms can be managed effectively and minimized.

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