



Chronic Intestinal Pseudo-obstruction in Children: An Overview

By: Paul E. Hyman, MD, Professor of Pediatrics, Louisiana State University and Chief, Pediatric Gastroenterology, Children's Hospital, New Orleans, LA

What is it?

Chronic intestinal pseudo-obstruction (CIP) is the name given to a number of rare disorders that cause impaired gastrointestinal motility (movement in the digestive tract). A diagnosis of CIP is based on symptoms that occur when the intestine is blocked and on clinical findings. In pseudo-obstruction, the symptoms are caused not by a surgically correctable tumor, twist, or ulcer in the bowel, but by a problem having to do with the strength or coordination of the contractions that move along the contents of the bowel. Children with CIP often complain of poor appetite, nausea, vomiting, heartburn, abdominal pain, abdominal distension, and constipation or diarrhea. Affected children may not grow or develop at the expected rate.

CIP is caused by several different nerve and muscle abnormalities of the digestive system. Some of these are inherited, and others are acquired before or after birth. Children with CIP have different complaints, which vary in severity, and respond differently to treatment, because there are different underlying causes of CIP.

Most affected children have symptoms beginning at birth. In many, the symptoms are so severe that the children cannot or will not eat, and special forms of nutritional support are needed from the beginning. Other children acquire CIP suddenly after a viral illness. In these children CIP may be caused by persistent inflammation in the nerves of the gastrointestinal tract, long after the infection is gone. Still others experience increasing digestive problems during their childhood and into adult life. Most children with CIP have problems related to their digestive system only. In some however, CIP is

complicated by urinary bladder malfunction, developmental delay, dysautonomia (abnormal involuntary nerve function) or some other birth defect. About 200 new cases of CIP are diagnosed in American children each year. Diagnosis is often delayed because CIP is a rare condition that mimics more common, surgically correctable conditions.

The majority of affected infants undergo many medical procedures before the diagnosis of CIP is considered. Sometimes, an absence of abnormality in laboratory or x-ray tests leads a doctor to conclude that symptoms are psychological in origin. Although the underlying disorder of impaired gastrointestinal motility in CIP is not *caused* by mental illness, concern does build when a diagnosis cannot be established for a seriously ill child, and a family's ability to cope can be stretched to its limits.

Diagnosis

There is no specific test to diagnose CIP. It is a diagnosis based on symptoms and findings after a physical examination, plus the proven absence of bowel obstruction. Once the diagnosis is considered, different tests can determine the causes of CIP. Sometimes, tubes are placed through the nose or the anus to measure the strength and patterns of contractions throughout the gastrointestinal tract. These tests are known as esophageal, gastric, antroduodenal, and colonic manometry. Other times, intestinal biopsies, or tissue samples, obtained at the time of surgery reveal the cause of CIP.

Treatment

About 10% of affected infants improve spontaneously over months or years. For the

remaining 90%, there are no cures for the neuromuscular diseases that cause CIP, but there are nutritional, medical, and surgical options available to promote normal growth and development.

Predigested liquid diets may be fed to children in a variety of ways, called enteral feeding. A nasogastric tube is a small plastic tube placed into the nasal passageway and down into the stomach. A gastrostomy is a small opening connecting the inside of the stomach to the outside of the abdominal wall. The gastrostomy eliminates the need for nasogastric tube feedings, and serves as a vent to relieve the pressure and pain caused by the backup of intestinal contents.

When the stomach itself is severely obstructed, children can be fed through a jejunostomy, a small opening created surgically from the middle part of the small intestine to the outside of the abdominal wall. In children who are unable to receive appropriate nutrition despite all attempts, enteral feeding is lifesaving.

As a last resort, total parenteral nutrition (TPN) involves surgically implanting a catheter, or plastic tube, into a large vein near the heart. A nutrient solution is infused by a small pump through the catheter directly into the bloodstream.

Surgery is often necessary to provide access to the route of nutritional support, either gastrostomy, jejunostomy, or central venous catheter placement of TPN.

The life-threatening aspects of CIP most often lie not in the disease itself, but in the complications that arise from the use of TPN. Severe blood infections and liver failure are the most common causes of death in children with CIP, and both of these are associated with long-term use of TPN.

Drugs that improve contractions in the gastrointestinal tract (promotility drugs) are useful in some children with CIP. For some children, pain management is a serious problem. New non-narcotic drugs and a variety of psychological strategies are used for treating chronic, deep abdominal pain.

Although the search for effective means of diagnosis and treatment can be elusive, the future for children severely affected with CIP is brightened by the evolving promise of cure with intestinal or multi-organ transplantation. Even better, scientists are working to replace damaged or absent nerve cells with stem cell transplants. Other scientists may be working on stem cell transplants of smooth muscle into the bowels.

Suggested IFFGD Reading

Hyman P, Di Lorenzo C. *Chronic Intestinal Pseudo-obstruction*. IFFGD. Fact Sheet No. 801.

Norton W, Di Lorenzo C. *Chronic Intestinal Pseudo-Obstruction*. IFFGD. Fact Sheet No. 843.

Opinions expressed are an author's own and not necessarily those of the International Foundation for Functional Gastrointestinal Disorders (IFFGD). IFFGD does not guarantee or endorse any product in this publication nor any claim made by an author and disclaims all liability relating thereto.

This article is in no way intended to replace the knowledge or diagnosis of your doctor. We advise seeing a physician whenever a health problem arises requiring an expert's care.

IFFGD is a nonprofit education and research organization. Our mission is to inform, assist, and support people affected by gastrointestinal disorders. For more information, or permission to reprint this article, write to IFFGD, 700 W. Virginia St., #201, Milwaukee, WI 53204. Call toll-free (In the U.S.): 888-964-2001 or 414-964-1799. Visit our websites at: www.iffgd.org or www.aboutkidsgo.org.