



Chronic Intestinal Pseudo-obstruction Syndrome in Children

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Introduction

Your child has been diagnosed as having intestinal pseudo-obstruction or some other form of chronic gastrointestinal motility disorder. This brochure has been written to help you understand these disorders and the effects they may have on your child. Because the symptoms and their severity are variable, medical, surgical, and nutritional management will depend on individual needs and resources. Therefore, we recommend that you discuss your child's problems with a healthcare professional.

What is gastrointestinal motility?

The food that you eat moves approximately 30 feet through the digestive system, starting from the esophagus through the stomach, small intestine, and ending in the colon. The movement of food is coordinated by the nerves and muscles that are part of the digestive tract. When food enters the gastrointestinal tract, the nerves tell the muscles to contract in movements that mix and propel the food, thereby making digestion possible. Motility refers to the muscle contractions within the walls of the digestive tract and the movement of the food within the digestive system.

What causes gastrointestinal motility disorders?

When the nerves or muscles in any portion of the digestive system do not function in a strong or coordinated fashion, the child develops symptoms. Common motility problems include heartburn and constipation. People experience heartburn when their stomach acid goes the wrong way, from the stomach up into the esophagus. Many children and adults also have problems with constipation because the contractions of the large intestine or the relaxation of the pelvic floor are poorly coordinated. Another relatively common problem is the delayed emptying of food from the stomach. This delay can result in chronic nausea, abdominal distention, and vomiting.

What is chronic intestinal pseudo-obstruction?

Chronic intestinal pseudo-obstruction (CIP) is the name given to a number of rare disorders that cause impaired gastrointestinal motility. A diagnosis of CIP is based on symptoms and body changes that occur when the intestine is blocked. In pseudo-obstruction, the symptoms are not caused by a surgically correctable tumor, twist, or ulcer in the bowel,

but rather by a problem having to do with the strength or coordination of the contractions that move contents along. Children with CIP often complain of poor appetite, nausea, vomiting, heartburn, abdominal pain, and constipation. As a result, normal growth and development may be interrupted.

Chronic intestinal pseudo-obstruction (CIP) is caused by one of 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, varying in severity and response to treatment.

Most affected children have symptoms starting at birth. In many, the symptoms are so severe that the child cannot or will not eat, and may require special forms of nutritional support. Other children acquire CIP suddenly after a viral illness. Still others experience increasing digestive difficulties during their childhood and into adult life. The majority of children with CIP have problems related to their digestive system only. In some, however, CIP is complicated by urinary bladder malfunction, developmental delay, or some other birth defect.

How many children are diagnosed with CIP?

About 200 new cases of CIP are diagnosed in American children each year. This suggests that CIP occurs in 1 in every 25,000 children. Diagnosis is often delayed because CIP is a rare condition that mimics more common surgically correctable conditions.

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

Diagnosis

There is no specific test to diagnosis CIP. It is a diagnosis based on symptoms and findings after a physical examination, plus the proven absence of a true bowel obstruction. Once the diagnosis is considered, testing sometimes helps in understanding the causes of CIP. Some of these tests are

known as manometry, which means measuring pressures. Manometry involves placing thin plastic tubes through the nose or anus to measure the strength and patterns of the contractions throughout the gastrointestinal tract. Manometry of the esophagus, stomach, small bowel, and colon are useful for determining the nature and extent of the abnormalities associated with CIP. Other times, intestinal biopsies, or tissue samples, obtained at the time of surgery reveal the cause of CIP when they are examined under the microscope.

Treatment options

About 10% of infants diagnosed with CIP improve spontaneously over months or years. For the remaining 90%, there are no known cures for the neuromuscular diseases that cause CIP. However, there are nutritional, medical, and surgical options available to promote normal growth and development.

Enteral feedings

Enteral feedings consist of predigested liquid diets which may be administered in a variety of ways. They may be taken as dietary supplements or as the sole form of nutritional support. They may be given in small, frequent feedings or drop by drop in a continuous infusion. An infusion of special formula requires the placement of a tube into the digestive tract.

A nasogastric tube (NG-tube) is a small plastic feeding tube placed through the nasal passageway and down into the stomach. Once in place, it is taped securely to the nose or the side of the face. The NG-tube is attached to plastic tubing which delivers formula at a predetermined rate. A nasogastric tube is not very practical for long-term management.

A gastrostomy (G-tube) is a small opening connecting the inside of the stomach to the outside of the abdominal wall. The gastrostomy eliminates the need for a nasogastric tube in children requiring very frequent or prolonged tube feedings. Moreover, in patients with abdominal distention and pain, the gastrostomy is used as a venting mechanism to relieve the pressure and pain caused by the backup of intestinal contents.

When the stomach is severely involved, and an NG- or a G-tube is not an option, children can be fed through a jejunostomy. A jejunostomy (J-tube) is a small opening created surgically from the middle part of the small intestine (jejunum) to the outside of the abdominal wall. Formula may be administered drop by drop throughout the day or continuously overnight.

Parenteral feedings

In children who are unable to digest food despite all attempts, parenteral feeding is lifesaving. Total parenteral nutrition (TPN) involves surgically implanting a catheter, or plastic tube, into a large vein near the heart. A solution containing nutrients may be infused via an intravenous pump through the catheter

directly into the bloodstream, for 12 to 24 hours daily. Surgery is necessary in order to provide access for the route of nutritional support when placing either a gastrostomy, jejunostomy, or central venous catheter for total parenteral nutrition.

Is CIP life threatening?

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

Drugs that improve contractions in the gastrointestinal tract (prokinetic drugs) are useful in some children with CIP. For others, pain management is a serious problem. Non-narcotic pain medicines and behavioral strategies are used by pediatric pain management specialists for the treatment of chronic and severe abdominal pain to improve the quality of life for children with CIP.

In conclusion

Although the search for an effective means of diagnosis and treatment can be elusive, the future for children severely affected by CIP is brightened by the evolving promise of a cure with an intestinal or multiorgan transplantation, pacing the intestine, and cloning stem cells to replace damaged or non-working nerves or muscle.

It is important to remember that each child and family requires individual assessment. This brochure is not meant to replace the diagnosis and treatment recommendations of your child's physician.

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