



# CdLS Foundation

Cornelia de Lange Syndrome Foundation, Inc.  
*Reaching Out, Providing Help, Giving Hope*

## GI DISTRESS — MALROTATION, VOLVULUS, BOWEL OBSTRUCTION, PANCREATITIS

*By Carol Potter*

*As Part II of a two-part review of GI issues in persons with CdLS, Dr. Carol Potter describes symptoms and treatments of malrotation, volvulus, bowel obstruction, and pancreatitis.*

### **Malrotation and Volvulus**

Symptoms of malrotation and volvulus may include vomiting, pain, a swollen abdomen, bloody or loose stools, rapid heart rate and breathing, little or no urine, and fever.

Malrotation occurs when the intestine or bowel does not fold or rotate properly in early fetal development. Malrotated intestines may “flop” around since they are not properly attached to the abdomen wall. This can result in the intestines twisting around one another. This is called volvulus. As this happens, the duodenum may be twisted shut which can induce vomiting with or without pain. If the intestines twist further, they may compress the blood supply. This can cause severe pain as the blood supply to the small intestine is cut off. Sometimes, the twist relaxes and the pain resolves. However, with each episode there is the chance that the blood vessel compression will last too long and the intestine will die. Unfortunately, the entire small intestine is usually involved, with the exception of the top few inches and the lower half the colon. When this much bowel is lost, most children require intravenous nutrition for the rest of their lives.

Malrotation is diagnosed by having the child swallow barium. It is important to track the barium as it travels through the first part of the small intestine to make sure the intestine passes correctly from right to left. Sometimes barium enemas are used to see the location of the beginning of the colon. Because malrotation can be a life-threatening emergency that most commonly occurs in childhood, pediatric radiologists are often more familiar with its subtle forms and more aggressive about watching barium long enough to make sure malrotation is not present. For this reason, we recommend that studies done to evaluate vomiting in children be done by pediatric radiologists whenever possible.

Malrotation is a medical emergency. If volvulus is not present, i.e. the bowel is not blocked from twisting and the blood supply is not being compressed, surgical correction is usually done within a few days. If vomiting is present and x-ray studies show that volvulus is present, surgical therapy should occur immediately. The surgeon will untwist the bowel and try to determine if any bowel has died. Once dead bowel is removed, the remaining intestine is attached to the back of the abdomen to prevent further volvulus.



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## **Bowel Obstruction**

Signs of obstruction or blockage include pain, vomiting (green or yellow bile), and distention of the abdomen. Unfortunately none of these can tell you definitively if the child does or does not have obstruction. They are, however, worrisome signs and should be investigated. Bowel obstruction simply means blockage of the gastrointestinal tract. This can occur anywhere from the mouth to the rectum. Congenital malformations (malformations present at birth) can cause bowel obstruction. Atresia, an absence of openings in areas of the intestine, is a common cause seen at birth.

Other congenital problems may show up later in life, including webs (thin membranes which stretch across part of the intestine). Malrotation and volvulus are also examples of congenital malformations that can show up at any age. Sometimes part of the GI tract has a duplication, or a second GI tract running beside it. This can be short or long and can sometimes cause blockage of the main intestinal tract.

Some bowel obstructions do not occur because of problems we are born with but problems we acquire during life. Adhesions, or bands of scar tissue can result after any surgery in the abdomen and can block the intestines. Funduplications (surgery done for gastroesophageal reflux) that are too tight can cause obstruction or blockage. Inflammation in the intestine or abdomen can also cause obstruction. Finally, severe constipation can cause a reversible obstruction of the GI tract.

## **Pancreatitis**

Pancreatitis means inflammation of the pancreas. There are many causes but they lead to a similar picture. The child may develop a severe abdominal pain, often accompanied by vomiting, over a relatively short period of time. The child's abdomen is often very tender and the child appears sick.

There are several causes of pancreatitis. Viruses can cause pancreatitis in children. A blow to the upper abdomen can also cause pancreatitis as the fragile ducts of the pancreas are damaged against the spine. Many medications, including some seizure medications and steroids, can also cause pancreatitis. Gallstones and congenital malformations of the bile ducts and pancreas can also predispose children to inflammation of the pancreas. The diagnosis of pancreatitis is made by blood tests. X-ray studies such as ultrasound, CAT scan and MRI may help determine the cause and severity of the pancreatitis.

The pancreas makes the enzymes we need to break down food. When the pancreas is in inflamed, these same enzymes start to digest the pancreas. If the process is mild, the child may get better with minimal or no intervention. More severe cases of pancreatitis often require admission to the hospital for IV fluids and pain control. A child may not be able to eat his/her regular diet until the condition improves. In a very severe case of pancreatitis, the child can become extremely ill and develop long term complications.



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The prognosis for children with pancreatitis is dependent on the cause of the inflammation and the severity of the illness.

***Children with CdLS are at risk for developing GI issues. Early recognition of the signs and symptoms of these conditions, quick medical action, and perseverance with medical professionals will equip parents and caregivers with the best defense against the development of more serious complications in their children.***