



## Malrotation (rotational anomalies, malrotation with volvulus)

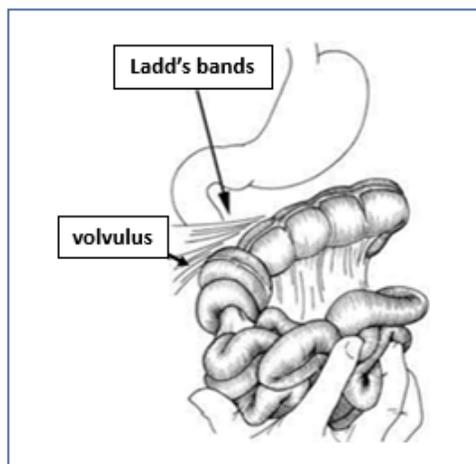
*Patient and family information, brought to you by the Education Committee of APSA*

### Overview - “What is it?”

The normal development of the intestines as a fetus (developing baby) in the first trimester of pregnancy, involves the intestines leaving the belly and growing outside the fetus’ belly. They return back into the belly after they mature and on their way back into the belly, they rotate to the normal position.

Malrotation is the failure of normal intestinal rotation and fixation during the development of the fetus. Malrotation happens when the intestine does not follow the normal position they are supposed to be inside the belly. When malrotation is present, there are two things that can give a child problems: volvulus and Ladd’s bands. (Figure 1)

- Volvulus is when the intestine twists upon itself, cutting off its own blood supply. Volvulus is dangerous because it can cut off the blood supply to most of the intestines causing the intestines, and sometimes the child, to die.
- Abnormal adhesions (Ladd’s bands) that can partially block the passage of contents within the intestines.



**Figure 1:** *Intestinal malrotation. The volvulus is the intestine that is twisted.*

*The Ladd’s bands are the adhesions that can block the intestine.*

*(Image credit: pedsurglibrary.com)*

Malrotation occurs in 1 in 200 to 500 of live births. Most patients have no problems or symptoms from the malrotation. Symptomatic malrotation occurs in approximately 1 in 6,000 of live births. Most patients have problems in the first months of life- up to 80% of babies with malrotation will be diagnosed the first month of life.

Malrotation is associated with other conditions including: Hirschsprung disease, small intestinal atresia, gastroschisis and omphalocele, congenital diaphragmatic hernias, cardiac heterotaxia

### Signs and Symptoms - “What symptoms will my child have?”

**Early signs** of malrotation depend of the problems it is causing. If there are abnormal bands partially blocking the intestines, the baby may have intermittent vomiting interfering with weight gain. If there is volvulus, the classic symptom is bilious (green or bright yellow) vomiting.

**Later signs/symptoms (more than a couple of hours)** of malrotation with volvulus include firm, distended and painful belly. There may also be blood in the stool, high heart rate, and low blood pressure. There may be decreased wet diapers. These symptoms may signal dead intestine.

### Diagnosis - “What tests are done to find out what my child has?”

**Physical examination:** Physical exam may include an enlarged belly and painful to touch. This is a concerning finding. Many times, the belly is not enlarged.

**Labs:** If the baby has lots of vomiting, blood tests will be sent to check levels of electrolytes (minerals) in the blood.

#### Imaging studies:

- Abdominal x-rays may be obtained in a child with vomiting, initially to look for different causes of vomiting.
- An upper gastrointestinal (UGI) contrast study is the test of choice if malrotation is suspected. The child is given contrast to drink. Several x-rays under a special machine are taken to follow the contrast through the stomach and into the first portion of the small intestine. This study shows whether the positioning of the intestines is normal or not.
- Ultrasound may be helpful to look for other causes of vomiting and belly pain such as intussusception.

**Conditions that mimic malrotation:** intestinal atresia, annular pancreas, meconium ileus, intussusception, Hirschsprung disease, gastroesophageal reflux, gastroenteritis.

## Treatment - “What will be done to make my child better?”

Treatment for malrotation depends on the symptoms the child has, the overall health including other medical problems, and the degree of malrotation.

- For example, in children with complex cardiac problems and malrotation but with NO symptoms related to malrotation, surgical intervention is not recommended. In these children, there are higher risks for surgery but potentially little benefit.
- In children who have symptoms of vomiting and inability to gain adequate weight or to advance volume of feeding, surgical repair is recommended on a scheduled (elective) basis.
- In patients with volvulus, the repair is performed on an emergency basis. The longer the intestines do not have blood flow, the higher risk it is for the intestines to die and the baby become sick.

### Malrotation without volvulus

**Medicine:** No medicine can make this condition better.

**Surgery:** This is usually a scheduled (elective) operation. The name of the procedure for malrotation is called the Ladd procedure, named after the surgeon who invented it. The abnormal bands causing partial obstruction are cut, the blood supply to the intestines is checked, and the appendix is removed. In malrotation, the appendix is found in the left side of the abdomen instead of the right side. The appendix is removed to prevent any confusion in the future if your child were to develop appendicitis. In this set of patients, Ladd procedure can be completed open or laparoscopic.

**Open:** A cut is made in the right upper part of the belly or through a vertical cut in the middle of the belly – depending on the size of the child.

**Laparoscopy:** Several small cuts (incisions) are made. Through one of the cuts, a video camera is placed. The surgery itself is done using small instruments placed through the other incisions. The usual number of incisions (cuts) for laparoscopic surgery can vary.

**Preoperative preparation:** Your child should shower or bathe the night before or the morning of surgery. No eating or drinking for 4-8 hours depending on your hospital’s recommendations. Prior to surgery, antibiotics may be given through the vein.

#### Postoperative care:

**Activity:** Typically, the child is encouraged to resume regular activity as soon as possible.

**Diet:** Oral feeding will resume once there is evidence the intestines have recovered from surgery. This is different from patient to patient.

**Medicines:** Your child may need any of the following:

- **Antibiotics:** To help prevent or treat an infection caused by bacteria.
- **Anti-nausea medicine:** To control vomiting (throwing up).

- **Pain medicine:** Pain medicine can include acetaminophen (Tylenol®), ibuprofen (Motrin®), or narcotics. These medicines can be given by vein or by mouth.

### **Malrotation with volvulus**

**Medicine:** Patients with malrotation with midgut volvulus will need an IV for fluids and medicines to prevent infection (antibiotics). If the patient is in pain, medications to help relieve discomfort may be needed.

**Surgery:** Volvulus is a *surgical emergency!* The procedure is usually done through a horizontal or vertical cut in the belly. First, the intestines are untwisted to restore blood flow. The intestines are assessed to see how much damage the volvulus caused. Sometimes, untwisting and restoring blood flow revives intestine. If the intestines are damaged beyond repair, they will need to be removed. After the damage to the intestines is assessed and taken care of, the Ladd procedure is performed.

**Preoperative preparation:** Patients need to be given fluids and antibiotics through the vein. Transfusions of blood products may be needed.

**Postoperative care:** Patients will remain on antibiotics after surgery. Patients will not be fed for a period of time after surgery until their intestine begins working again. If there is a question on whether segments of intestine can recover, the patient may be taken for another operation soon after the first, for additional evaluations prior to closing of the abdomen. Some patients may need to be in the intensive care unit (ICU) or have nutrition through a special IV called total parental nutrition (TPN).

**Risks:** Bleeding, infection, injury to abdominal structures such as intestine. If the volvulus is present for a long time and a large portion of intestine is unable to be salvageable, patients are at risk of a death or a disorder call “short gut syndrome.”

**Benefits:** Surgical intervention is required to improve symptoms of malrotation with or without volvulus. In cases where there is suspected volvulus, surgery may be lifesaving.

### **Home Care - “What do I need to do once my child goes home?”**

**Diet:** Will be based on how much intestine remains – Some patients may need TPN. Otherwise, patients will return to a regular oral diet.

**Activity:** Normal infant/pediatric activity

**Wound care:** Keep the incisions dry for 48-72 hours after surgery. The incisions can then be washed with soap and water gently while refraining from submerging incisions in the water for 1-2 weeks after the surgery.

**Medicines:** Patients may need pain medications such as Tylenol and Ibuprofen.

**What to call the doctor for:** Green vomit, not tolerating feedings, fever (>101 F), redness around the incision, or bloody or yellow/green drainage from the surgical incisions.

**Follow up care:** Surgery follow-up in 1-2 weeks. Routine pediatrician follow-up.

### **Long Term Outcomes - “Are there future conditions to worry about?”**

**Malrotation without volvulus:** long-term outcomes are excellent. There is a 10-15% risk of intestinal obstruction in the child’s lifetime. This complication is usually suspected if green or bright yellow vomiting, belly pain happens.

**Malrotation with volvulus:** Long-term outcomes where intestines were removed are dependent upon the length of intestines remaining after surgery. Children with no or little bowel removed will have normal intestinal function. Children with a small amount of intestine left are at risk for short bowel syndrome. These patients require close follow up with surgery and gastroenterology (specialists of intestinal function).

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