Gastroschisis

What is Gastroschisis?

Gastroschisis is a defect in the development of the abdominal (belly) wall of the fetus. This opening in the abdominal wall, usually to the right of the umbilicus (belly button), allows variable amounts of intestine to protrude freely into the amniotic fluid. This defect occurs in about 1 in every 3,000 live births.

How is Gastroschisis Diagnosed?

Gastroschisis is usually detected by ultrasonography in the prenatal period. Early detection allows for proper preparation of the parents, obstetrician or perinatologist, neonatologist, and pediatric surgeon.

Are there Associated Anomalies?

Fortunately, additional defects are not common with gastroschisis, and those that do appear are usually limited to the intestine. These defects are usually stenosis (narrowing), atresia (absence of a section), or shortened overall bowel length. Rarely, in cases with a particularly small abdominal wall defect, the blood supply of the entire intestine may be compromised, leading to loss of nearly all of the bowel and the condition called short bowel syndrome.

What is the Treatment for Gastroschisis?

Delivery is generally arranged by the obstetrician or perinatologist at around 37 weeks of gestation. A scheduled delivery allows the neonatologist and surgeon to prepare everything the baby will need in advance.

After the baby is born, the exposed bowel is immediately protected by sterile saline soaked dressings covered by a plastic wrap, minimizing infection and heat loss. A naso-gastric tube (tube from the nose into the stomach) will be placed to keep air and secretions from going into the intestine and to prevent vomiting. An intravenous line will be placed and antibiotics started. Although, associated anomalies are unusual, a pediatric cardiologist may be consulted.

There are two methods for restoring the intestine to the abdominal cavity and closing the defect — immediate repair or gravity reduction.

- **Immediate Repair**
  - If no other problems are found which would delay surgery, the baby is taken to the operating room and placed under general anesthesia.
First, a central line, called a broviac may be placed. This catheter is tunneled from the chest wall to the vein to minimize the risk of infection. This catheter allows intravenous nutrition to be given until the baby can feed normally which can take a month or more in some cases.

The intestine is gently placed back into the abdominal cavity and the defect closed.
- Occasionally, the intestine cannot be fully reduced back into the relatively small abdominal cavity because it causes too much pressure.
- The intestine pushes up on the diaphragm (muscle separating the chest and abdomen) and can cause respiratory distress and blood flow to the intestine and kidneys can be compromised, as can the return blood flow from the lower body.
- If this is the case, the surgeon will place a sterile sac (silo) over the intestine to allow it to gradually fall back in to the abdomen with the help of gravity and some external pressure as we tighten the sac.
- This allows the abdominal cavity to slowly enlarge and prevents compression of the lungs and organs. When the intestine is nearly all back into the abdomen, in approximately 4-7 days, the baby is taken back to the operating room and the abdominal wall is closed.

If there is a segment of intestine which is narrowed or missing, causing a blockage of the intestine, immediate repair or more often, a delayed repair will need to be performed.

Perforated (ruptured) intestine may need to be brought to the skin surface which can complicate the repair and will require additional operations.

Gravity Reduction
- Soon after birth, the surgeon gently places the intestines into a clear, sterile silicon sac. The sac has a spring gasket that slips into the abdominal wall defect.
- Next, the bag is suspended from the baby’s bed. While the swelling in the bowel decreases and the abdominal wall gradually stretches, the intestines gradually drop back into the abdominal cavity.
  - This process takes about five days.
- Placement of the bag and the ensuing gravity reduction take place in the NICU.
- Little sedation is required.
- After the bowel has dropped back into the abdomen, the baby is taken to the operating room and the defect is closed.
- Whenever possible, the skin is closed around the umbilical stump to approximate the look of a normal belly button.
- A central line (broviac) may also placed as described above.

Patience is needed after the surgery is completed. The intestine that was exposed to the amniotic fluid may be quite swollen and it may take three to six weeks for bowel function to return. Then it will take time before the baby is able to take enough oral nutrition for growth.

What are Possible Complications?

- Any baby undergoing general anesthesia and abdominal surgery has the risks of the anesthesia, bleeding, infection, and intestinal obstruction from scar tissue.
- The primary risk of a central line is infection. As pediatric surgeons, we place hundreds of such lines each year for this purpose as well as for the administration of medicine. If these lines get infected, antibiotics are used for treatment. In some cases the line must be removed. There can be long term metabolic effects of intravenous nutrition on the liver. These will be explained to you by the neonatologist.
The major complications of the abdominal wall closure are bleeding, infection, bowel injury, and abdominal compression resulting in respiratory compromise (in addition to any intrinsic respiratory problems), bowel compromise, and low blood pressure, all of which may require re-operation.

Bowel obstruction from scarring within the abdomen is less than 3%.

Gastroesophageal reflux is not unusual in these babies. Most can be managed medically but occasionally surgery is necessary.

Incisional hernias (weakening of the incision) with separation of the abdominal wall muscle occurs in approximately 10% and this will require re-operation to repair the defect later on.

Because of the protruded intestine, the bowel is not usually in a normal position (malrotation).

While malrotation in association with abdominal wall defects rarely causes a problem such as twisting of the intestine (volvulus), there may be other problems.

For example, if the child develops appendicitis in the future, the appendix and thus the pain, may appear anywhere in the abdomen, not just in the right lower abdomen. Parents should always mention the history of gastroschisis to the child's caregivers.

What is the Expected Outcome for babies with Gastroschisis?

Most babies with gastroschisis do quite well. Without associated anomalies, one can expect a 95-97% survival.

Disclaimer: Your child's condition is unique. The information contained on this web site is not intended to substitute for advice from a doctor or nurse. If you are unsure about any aspect of your patient's care, please contact us at 303-839-6001, or talk to your pediatrician.

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