

Questions and Answers About:

Gastroschisis

October 2010

What Is Gastroschisis?

Gastroschisis occurs when a hole develops in the fetus' abdominal wall. Through this hole the intestines go out of the abdomen and outside of the body. During pregnancy, the intestines float in the amniotic fluid, which irritates the intestinal tissue. At birth, the intestines can be seen outside of the abdomen (Figure 1). Gastroschisis usually develops on the right side of the umbilical cord early in the pregnancy, by 6-10 weeks after conception. As shown in Figure 1, the intestines often appear swollen and matted together.

Why Is Gastroschisis a Problem?

Other than the obvious fact that the intestines must be surgically placed into the abdomen, exposure to the amniotic fluid interferes with normal intestinal development and function. After



Figure 1. Picture of gastroschisis with intestine in a matted mass. The infant's head is to the right. Photo courtesy of David Rustad, MD.

exposure to amniotic fluid, the intestines do not function properly and often take many weeks to develop the rhythmic contractions that move food through the intestinal tract. Because the intestines do not develop in their normal position, they can also become kinked or blocked or lose part of their blood supply prior to birth. At least one and sometimes several operations are required to place the intestines into the abdominal cavity and

fix any intestinal blockages.

How Often Does Gastroschisis Occur?

The frequency has increased over the past 20 years from 1 in every 5,000 to 1 in every 2-3,000 live births. Gastroschisis occurs more often in young women, especially those less than 20 years of age. Women who smoke and are quite thin also have increased risk. We do not know why these factors increase the risk of having a baby with gastroschisis.

Diagnosis

Fetal ultrasound can diagnose gastroschisis as early as 14 weeks after conception. The ultrasound shows free-floating loops of intestine in the amniotic fluid with no surrounding tissue membrane. The obstetrician may also obtain several ultrasounds during pregnancy to monitor fetal growth and intestinal development. Fetuses with gastroschisis often grow more slowly than babies without this problem.

Maternal Serum Alpha-Fetoprotein (MSAFP)

MSAFP is part of the Triple, Quad, or Penta screens used to look for Down syndrome and other birth defects. Prolonged, direct exposure of the fetal intestines to the amniotic fluid elevates a woman's MSAFP level. This often prompts the obstetrician to obtain a detailed fetal ultrasound, which identifies the gastroschisis defect. Although the screens are used to help identify Down syndrome, there is no connection between Down syndrome and gastroschisis. The MSAFP level is low with Down syndrome and elevated with gastroschisis. Women whose fetuses have spina bifida and related birth defects also have elevated MSAFP. However, there is no connection between spina bifida and gastroschisis.

During Pregnancy

Obstetricians often monitor pregnancies more closely than usual if the fetus has gastroschisis. The fetal intestines can become dilated (increased in size), and in extreme cases the fetus requires an early delivery before labor naturally

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starts. However, for most babies, mild to moderate intestinal dilation does not require early delivery.

Babies with gastroschisis often grow less quickly than other fetuses. Decreased growth requires careful assessment by the obstetrician because it can cause fetal problems during labor and require a Cesarean section. Babies with gastroschisis deliver at an average gestational age of 36 weeks.

Natural or Cesarean Birth?

Cesarian delivery does not usually improve the outcome of gastroschisis patients. There are special situations in which cesarean delivery is indicated. Examples would be when the baby is not tolerating labor or if there is an unusually severe problem with the intestines as a result of gastroschisis.

Will My Baby Survive?

Of our past 191 patients (since 1990), all have survived to go home from the hospital. However, they can have a long, complicated hospital course. About 30% of gastroschisis babies will be re-admitted to the hospital during their first 12 months after birth. About one-third of these re-admissions are for intestinal problems and the rest are for lung problems.



Figures 2 a-c. These three figures starting in upper left and moving clockwise show the same patient before reduction of the intestines into the abdomen, after reduction, and after closure of the hole in the abdominal wall. Photo courtesy of Pediatric Surgical Associates, P.A.

How is Gastroschisis Treated?

Treatment consists of two parts: returning the intestines to the abdomen and encouraging intestinal function. A single surgical procedure completely repairs most cases of gastroschisis.

Surgeons return the intestines into the abdominal cavity, and sew up the hole in the abdominal wall (Figures 2 a-c).

What Is a "Silo"

In about 50% of patients the intestine won't easily fit into the abdomen at the first surgery.



Figure 3. Photograph of "silo" enclosing the intestine. The "silo" is gently squeezed to push the intestine back into the abdomen over several days. Photo courtesy of David Rustad, MD.

These patients require a "silo", a plastic covering (similar to a sock) attached to the abdominal wall and containing the exposed intestine (see Figure 3). Over about 10 days, the surgeons gently push the intestines into the abdomen as the "silo" is rolled down, like a tube of toothpaste. At a second surgery the surgeon removes the "silo" and closes the abdominal wall. A "silo" increases the hospital stay by 10-14 days.

Complications

Some babies with gastroschisis also have atresias (blockages of the intestine). These occur in about 15% of cases. In about 3% of blockages, the intestine develops a hole that allows the intestinal contents to leak into the abdominal cavity. In another 3% of cases, the intestinal blood supply becomes injured during pregnancy due to the intestines' unusual location outside of the abdomen. These complications may require multiple surgeries and the creation of an ileostomy. Surgeons create an ileostomy by sewing a loop of intestine to the abdominal wall. The baby then passes stool through the ileostomy into a bag on the abdominal wall. An ileostomy is usually temporary for about 6-9 months and gives time for the intestines to heal.

Aside from surgical treatment of the gastroschisis, the intestines may require weeks to months

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to develop normal function. Prolonged feeding problems and slow weight gain most often occur when an atresia, intestinal leak, or poor intestinal blood flow complicates gastroschisis. Until the intestinal function normalizes, babies must be fed by vein. Because many babies with gastroschisis can't eat normally for many weeks or months, they often remain in the hospital for a long time receiving these intravenous feedings.

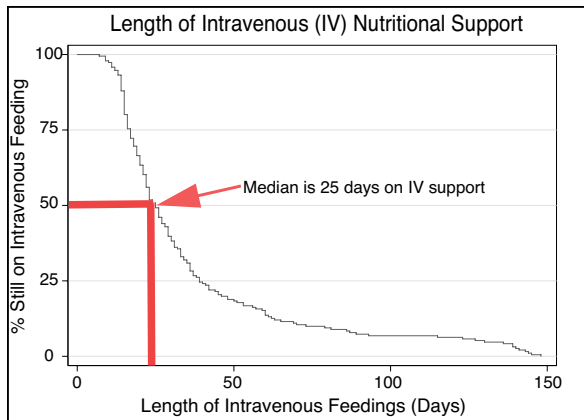


Figure 4. The Y-axis represents the percentage of patients who were still receiving IV nutrition at the corresponding day. The X-axis represents the total number of IV days. The red lines drawn on the graph represent the usual (median) number of days a baby with gastroschisis will receive IV nutrition (25 days).

Intravenous Feedings

For some babies, establishing intestinal feedings requires many weeks and therefore they require many weeks of intravenous feedings. Babies with gastroschisis usually require intravenous nutrition for about 25 days (Figure 4).

At the first surgery, most gastroschisis patients receive a special intravenous line. This is called a Broviac catheter and is inserted through the skin of the chest, tunneled under the skin up to the vessels of the neck where the catheter is inserted into a vein, and threaded down into a large vein near the heart. The intravenous feedings, called hyperalimentation or total parenteral nutrition (TPN) contain very concentrated solutions of protein, sugar, minerals, fat and vitamins. These concentrated solutions can irritate small veins with low blood flow. Using a large vein, close to the heart with a high blood flow, reduces vein irritation and the number of times that the intravenous catheter must be changed.

How Long Will My Baby Be in the Hospital?

On average, babies with gastroschisis spend 32 days (the median) in the hospital, although this varies widely depending on how difficult it is to establish intestinal function (Figure 5). About 90%

of patients with gastroschisis have gone home by 3 months of age. The length of the hospital stay depends on the degree of intestinal problems and the degree of prematurity.

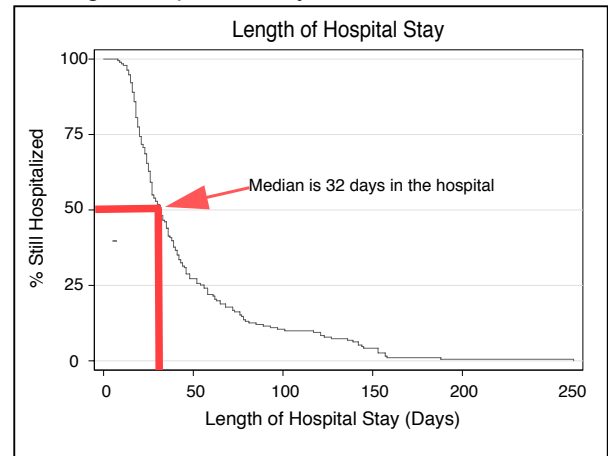


Figure 5. The Y-axis represents the percentage of patients who were still hospitalized at the corresponding day. The X-axis represents the number of total days spent in the hospital. The red lines drawn on the graph represent the usual (median) number of days a baby with gastroschisis will stay in the hospital (32 days).

How Long Will My Baby Be on a Ventilator?

Immediately after surgery, infants with gastroschisis need a ventilator in order to breathe. The average length of time a baby with gastroschisis spends on a ventilator is 4 to 5 days (Figure 6).

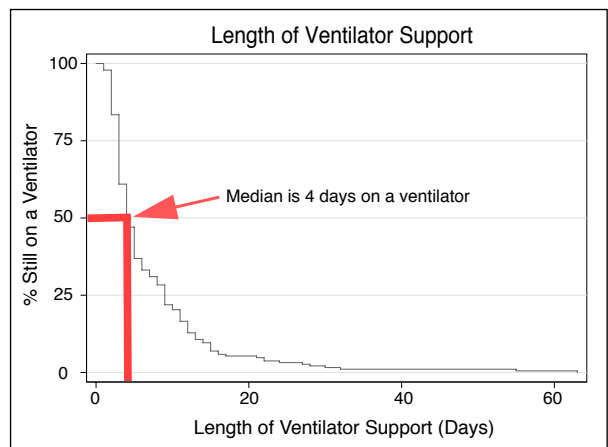


Figure 6. The Y-axis shows the number of patients still on a ventilator. The X-axis shows the total number of days spent on a ventilator. The red lines on the graph show the usual (median) number of days a baby with gastroschisis is on the ventilator (4 days).

Risk of Another Child with Gastroschisis

Fewer than 1% of our patients with gastroschisis have had the same mother. Although recurrence of gastroschisis in a family is rare, all future pregnancies should be monitored by ultrasound for gastroschisis. Research shows that gastroschisis can also occur in half-siblings with different fathers, although no pattern of heredity has been identified.

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After the Hospital

Most babies with gastroschisis do well following discharge. However, many continue to grow slowly for the first few years. By 3 years of age, most have caught up with their peers and are well above the 10th percentile on the growth curve. Some babies with gastroschisis require prolonged intravenous nutrition (TPN) either in the hospital or at home. All gastroschisis patients are also more prone to develop intestinal problems in the future due to kinking of the intestine. If your child develops severe abdominal pain or has other sudden intestinal symptoms, you should have a doctor examine him/her promptly.

If there has been an intestinal resection or if intestine had to be removed at surgery, feeding and growth issues may persist. Your primary doctor will monitor your baby's growth.

We are currently conducting a study of the long-term outcome of gastroschisis patients. So far, our results are encouraging and show that most babies develop normally, although they may grow slowly in the first few years. You may be asked to participate in this study after your baby is discharged from the hospital.

If you have questions at any time, please contact us (Minneapolis Campus: 612-813-6288 or 612-813-6295. St. Paul Campus: 651-220-6260).