Gastroschisis  Sequelae
and Management

Mary Finn
Gillian Lieberman, MD
Primary Care Radiology
Beth Israel Deaconess Medical Center
Harvard Medical School
April 2014
Outline

I. Definition and Epidemiology
II. Most Frequent Complications
III. Radiologic Investigations
IV. Index Patient Introduction
V. Companion Patient
VI. Take-Home Points
Gastroschisis

Represents a herniation of abdominal contents through a paramedian full-thickness abdominal fusion defect. The abdominal herniation is usually to the right of the umbilical cord.

No genetic association exists!
Gastroschisis looks similar to an Omphalocele.

An omphalocele, however, is a birth defect in which the infant's intestine or other abdominal organs protrude through the navel and are covered with parietal peritoneum.

Approximately 15% of live-born infants with omphalocele have chromosomal abnormalities. Severe malformations are present, such as cardiac anomalies (50%) and neural tube defect (40%).
Gastroschisis vs Omphalocele
Epidemiology

- Gastroschisis occurs in approximately one in every 2,000 live births and the incidence appears to be increasing.
- The cause of gastroschisis is unknown.
- This condition does not run in families.
- Most infants with gastroschisis do not have chromosomal or genetic syndromes, but sometimes may result in low birth weight, preterm delivery, and stillbirth.
- Gastroschisis has a strong association with low maternal age.
Most Frequent Complications of Gastrochisis:

• **CHOLESTASIS** (secondary to long term total parenteral nutrition)

• **MALROTATION, MIDGUT VOLVULUS, HYPOPERISTALSIS, GASTROESOPHOGEAL REFLUX (GER), AND ASPIRATION PNEUMONIA**

• **INTESTINAL ATRESIA/STENOSIS, SEPSIS, AND NECROTIZING ENTEROCOLITIS (NEC).**
Cholestasis

• Parenteral nutrition is a life-saving treatment for patients who have acute and chronic intestinal failure.

• Severe cholestasis induced by total parental nutrition (due to ‘excessive’ infusion of fat, carbohydrates and amino acids that precipitate) is characterized by bile duct regeneration, portal inflammation, and fibrosis.

• Its progression could be very rapid, and in some patients liver cirrhosis may develop within a few months.
• **Intestinal Atresia** is seen in approximately 20% of newborns with gastroschisis.

• The development of intestinal atresia/stenosis occurs secondary to torsion and volvulus of the exteriorized bowel, causing a disruption of mesenteric vessels and blood flow to the affected intestine.

• Bowel resection and anastomosis are frequently required and this places the infant at risk for **Short Bowel Syndrome**.
Radiologic Investigations:

- Plain Imaging
- Contrast studies (still X-rays or fluoroscopic X-ray images)
- Sonography
- CT in children less than 9 years old
- MR Enterography in children older than 9 years
Let’s Meet our Patient

Patient J. is a 7-year-old male (former 35 week gestation infant) with a history of gastroschisis with proximal atresia and subsequent resection of 15 cm distal jejunum d/t multiple perforations. He is S/P Roux-en-Y gastrojejunostomy, and STEP procedure times two. He is currently in bowel continuity, and has a CVC that is not currently being used for PN or IVF. His abdomen is distended and he has a history of poor weight gain.
Treatment and Medications

• Parenteral nutrition (PN) and intralipid from birth until July 2013.
• Eventually weaned off of PN but remained on IVF until January 2014.
• Currently eats a regular diet, without restriction.
• Has been on IV Metronidazole twice a day x 3 years for small bowel bacterial overgrowth symptoms.
• Takes Ergocalciferol 50,000 unit weekly.
• Scheduled to perform a DEXA scan and bone age evaluation.
• Fluoroscopy was performed as J. is a new patient at this hospital.
Fluoroscopy is a study of moving body structures--similar to an X-ray "movie." A continuous X-ray beam is passed through the body part being examined. The beam is transmitted to a TV-like monitor so that the body part and its motion can be seen in detail.

http://www.radiology-equipment.com/fluroscopy.cfm
Fluoroscopy lasted 1 hour and 15 minutes, a marked improvement from 18-hour-long previous investigation. The patient drank a barium swallow solution that coated and outlined the walls of his gastrointestinal tract.
A series of X-rays taken of the stomach and small and large bowel demonstrate marked small bowel dilatation and abnormal configuration. No obstruction was identified.
The patient had undergone the Roux-en-Y and STEP Procedures.

Contrast backflows to the blind end of the small intestine that was surgically divided off.
Roux-en-Y was performed because of proximal jejunal atresia and multiple perforations.
The **STEP Procedure** (Serial Transverse Enteroplasty) is a surgical technique for treatment of **Short Bowel Syndrome**, a condition in which nutrients are not properly absorbed because a large part of the small intestine is missing or has been surgically removed.

Companion Patient

• Patient K. is an 8-year-old girl born with gastroschisis and neonatal midgut volvulus.
• K. underwent a multivisceral transplant within three months of birth.
• Most recently she had mesenteric venous thrombosis, as well as renal lesions, for which she underwent extensive work-up, without finding any clear etiology.
• Her problem list is quite extensive and includes recurrent HSV stomatitis, azygos thrombosis, left lower lobe pulmonary embolism, portal vein and SMV thrombosis while on anticoagulation therapy.
Normal Anatomy of Aorta and Branches
3D Reconstruction of K.’s Post Transplant Arteries
Thrombosed Portal Vein
Massive Collateral Anastomoses Arose
Follow-Up

• Both J. and K. are currently doing well.
• They are able to stay with their families and lead a relatively productive life.
• The follow-up visits will include assessment of adequate growth and weight gain as well as developmental milestones.
• K.’s transplanted viscera are functioning properly. She will need to get laboratory tests done every three months for the rest of her life.
Take-Home Points

• Gastroscisis is a stand-alone congenital defect. Infants with Omphalocele have greater associated morbidity due to a higher incidence of congenital anomalies.

• However, there are numerous complications that may occur secondary to the evisceration of the intestines in gastroscisis, requiring long-term follow-up.

• With the advances seen in neonatal medicine (including surgical techniques, parenteral nutrition, respiratory support, and control of infection) these infants may go on to lead healthy and productive lives.
Special Thank You to

Neda Sedora-Roman, MD, BIDMC

Jamie Frost, DO, Boston Children’s Hospital
References

• http://www.cdc.gov/ncbddd/birthdefects/gastroschisis.html
• http://www.lucinafoundation.org/birthdefects-gastroschisis.html
• www.radiology-equipment.com/fluroscopy.cfm
• http://www.ncbi.nlm.nih.gov/pubmed/18242499
• http://www.ncbi.nlm.nih.gov/pubmed/18655097