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Briefly Legal: Should Midgut Volvulus Have Been Suspected in this Baby?

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A term baby was delivered by normal spontaneous vaginal delivery to a G_6P_3 with an uncomplicated pregnancy. Her cervical and rectal cultures for GBS were negative. Prenatal ultrasounds revealed exuberant fetal growth but no anomalies. There was no evidence of polyhydramnios or dilatation of the fetal bowel. At term, the mother went into spontaneous labor and delivered a 4,395-g female, who received Apgar Scores of 8 and 9 at 1 and 5 minutes, respectively. The newborn had no medical issues and was discharged home 48 hours after delivery.

"One month later, the baby suddenly developed poor feeding. The father describes the abdomen as becoming very hard and pale above the umbilicus and blue below it. He immediately brought the baby to the emergency department at a local hospital, where a physical examination revealed the baby to be non-responsive with weak pulses and a heart rate of 111 bpm. The temperature was 98 °F, and there was prolonged capillary refill. Spontaneous breathing was labored. The abdomen was markedly distended and firm. The skin above the umbilicus was pale and mottled from the waist down. Blood pressures were not attempted. A nasogastric tube was inserted with the return of 285 ml of bilious fluid, including some bright red blood."

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A complete blood count showed a hematocrit of 36%, a white blood count of 30,000, a normal platelet count, normal electrolytes except for a potassium level of 7.4 mEq/L, and elevated liver function tests. Following the successful resuscitation and the return to a normal heart rate, the baby was sent to the Radiology Department for chest and abdominal radiographs, abdominal ultrasound, and CT of the chest, abdomen, and pelvis. Upon return from the Radiology Department, her temperature was 92.7 °F. The radiologic studies were consistent with a small bowel obstruction. The ED physicians consulted the on-call surgeon, who was unable to come up with a diagnosis. He did not come to the hospital to see the baby but advised the ED team to continue observation and to send the baby to the Pediatric Intensive Care Unit (PICU). After blood cultures were taken, the baby was started on broad-spectrum antibiotics.

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After being admitted to the PICU, the baby received more boluses of fluid and multiple vasopressors to normalize her unstable blood pressure. On admission to the PICU, an arterial blood gas showed a pH of 6.8 and a base deficit of 30—a severe metabolic acidosis. A repeat hematocrit was 14%, for which blood transfusions were provided. She developed multiorgan failure with anuria, disseminated intravascular coagulopathy (DIC), and sustained hypotension. The following day, the baby was transported to a neighboring medical facility for dialysis. The admitting physicians, however, were concerned about her intestinal status and immediately set in motion preparations for exploratory laparotomy

within 2 hours of admission. At laparotomy, a midgut volvulus was found, with 70% of the intestine being necrotic. Following the surgery, the infant suffered through a prolonged, stormy postoperative course involving seizures, central line infections, and bacterial endocarditis. A brain MRI showed multiple foci of restricted diffusion one month after the surgery. At five months of age, the child was discharged from the hospital with diagnoses of small bowel syndrome and difficult-to-control seizures. Subsequently, she had multiple admissions due to problems with abdominal adhesions, complications from her central line, and intractable seizures. Follow-up at seven years of age revealed that she has severe cerebral palsy, developmental delays, short gut syndrome, cholestasis, and ongoing seizures. She is entirely dependent on parenteral nutrition and will require 24-hour care for the rest of her life.

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The Emergency Department, PICU physicians, and the hospital were sued. The trial resulted in a sizeable verdict for the plaintiff.

Allegations by Plaintiff:

Against the Emergency Department:

- A prolonged period in ED before evaluating blood pressure
- Failure to consider midgut volvulus or any other intraabdominal condition in the differential diagnosis
- Failure to keep baby euthermic
- Failure to perform a timely exploratory laparotomy
- Failure to stabilize in the ED
- Transporting infant to Radiology Dept before stabilizing
- Failure to order upper gastrointestinal examination

Against the Pediatric Intensive Care Unit physicians, including surgeon:

- Failure to consider midgut volvulus or other intraabdominal conditions in the differential diagnosis
- Failure to order upper gastrointestinal examination
- Failure to perform a timely exploratory laparotomy
- Failure to timely transport to another facility when the surgeon declined to do an exploratory laparotomy

Positions taken by the defense:

- Signs of volvulus were not present.
- Her abdominal distension was secondary to an ileus from sepsis or trauma.
- She was not stable enough to take to surgery.
- She was not stable enough to do an upper gastrointestinal series.
- Concern for aspiration if UGI were performed.
- Her GI and neurologic sequelae were not preventable; they were secondary to a lack of blood flow (ischemia) during her cardiac arrest.
- The differential diagnosis included sepsis, liver protein allergy syndrome, abdominal distension from postcardiac arrest, and volvulus.
- Everyone met the standard of care.

Discussion:

General:

Rotational anomalies of the gastrointestinal system occur due to an arrest of normal rotation and fixation of the intestinal tract in the abdomen. In the normal course of embryonic development, between 6 and 12 weeks of gestation, the gastrointestinal tract undergoes two definable, independent, 270-degree counterclockwise rotations. The first rotation involves the duodenojejunal junction around the axis of the superior mesenteric artery, and the second involves the ileocolic junction around the same axis. Under normal circumstances, the bowel does not twist, obstruct, or compromise its blood supply. Malrotation, however, does occur in about 1 in 6,000 live births. Sixty-two percent of these children have an associated anomaly, most being GI-related, but there may be concurrent congenital heart disease, especially heterotaxy syndrome and Cornelia de Lange syndrome.

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Intestinal malrotation is prone to midgut volvulus, thereby causing intestinal obstruction, leading to ischemia and necrosis and requiring emergent surgical intervention. Rare instances of in-utero midgut volvulus have been reported in the literature and carry a high mortality, given the difficulty in establishing a diagnosis prior to the development of signs of intestinal ischemia and necrosis. However, more recent advancements in imaging and increasing awareness have made it possible to diagnose malrotation prenatally, raising the question of optimal timing of delivery, especially in cases of prenatally diagnosed midgut volvulus. Failing a prenatal diagnosis, the newborn may present

at birth with abdominal distension and the need for emergency care and operation. Malrotation may exist without volvulus or compromise. Non-emergent malrotation may present in older children or adults; they are found during an evaluation for intermittent chronic pain or vague GI symptoms or as an incidental finding on upper gastrointestinal contrast studies, about 2/1000.

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Prenatal Detection:

Advances in ultrasound imaging have facilitated the intrauterine diagnosis of malrotation, which demands deciding on the optimal timing of delivery, especially in cases of prenatally diagnosed midgut volvulus. Here, the risks of premature birth must be weighed against the risks of fetal intestinal ischemia and potential fetal demise. Abnormal FHR patterns may assist in the diagnosis of systemic involvement. Ultrasonographic signs can be direct and specific ('whirlpool sign,' 'twining sign,' 'coffee bean sign') or indirect and non-specific (abdominal mass, dilated bowel loops, pseudocysts, ascites, absence of abdominal peristalsis, polyhydramnios, echogenic bowel). Early suspicion of intestinal volvulus allows the clinician to refer the patient to a tertiary center for confirmation of the diagnosis and pursue appropriate follow-up.

Presentation:

Several variations of malrotation exist, which may present with different clinical pictures. An acute presentation may be due to duodenal obstruction. Here, the patient usually presents with vomiting, especially bilious, hemodynamic instability from hypovolemia and/or septic shock. Physical examination includes abdominal distension and tenderness, and at times, hematochezia secondary to bowel ischemia and possible necrosis—more than half of the patients with duodenal obstruction present in the first month of life. The prognosis of babies with intestinal volvulus depends on the length of the segment involved, the level of intestinal obstruction, the presence of meconium peritonitis, and the gestational age at birth.

"The diagnosis of intestinal malrotation should be suspected in any infant who presents with bilious emesis, acute abdominal distension or tenderness, or evidence of duodenal obstruction on a radiograph. Depending on the completeness and duration of the obstruction, hemodynamic deterioration soon follows. Midgut volvulus is a life-threatening condition that requires emergent evaluation and intervention."

The diagnosis of intestinal malrotation should be suspected in any infant who presents with bilious emesis, acute abdominal distension or tenderness, or evidence of duodenal obstruction on a radiograph. Depending on the completeness and duration of the obstruction, hemodynamic deterioration soon follows. Midgut volvulus is a life-threatening condition that requires emergent evaluation and intervention. No additional evaluation is warranted if the patient has volvulus with signs of systemic decompensation. The patient should be rapidly resuscitated and immediately taken to surgery for exploration. Midgut volvulus with obstruction is one of the few pediatric emergencies in which the need for emergency surgery takes precedence over the need for hemodynamic stability. Resuscitation will have to be continued during surgery. As the operation gets underway, intravenous fluids continue, bladder and stomach catheters can be placed, blood is drawn for type and cross-match, and broad-spectrum antibiotics are administered. Time is critical. In patients with malrotation without an obstruction, urgency is somewhat less.

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Older children may present less dramatically, if at all. Symptoms may be present over hours or days or as chronic intermittent pain over weeks, months, or years. Intermittent vomiting, chronic diarrhea, malabsorption, or failure to thrive comprise other potential presenting symptoms in the pediatric population.

Images for Diagnosis:

If the pediatric patient is hemodynamically stable, the diagnosis should be confirmed by radiologic evaluation, typically beginning with **plain radiographs**. These are not diagnostic but are valuable as a screening tool to exclude perforation and expedite surgical exploration. The plain radiograph may show a nasogastric tube extending into an abnormally positioned duodenum or slow a "double-bubble," signifying duodenal obstruction. Partial obstruction of the duodenum causes distension of the stomach and the first part of the duodenum. Once perforation is excluded by left lateral decubitus or cross-table lateral films, plain radiographs are followed by an **upper gastrointestinal (UGI) contrast** series to visualize the duodenum. This is specific and sensitive for the diagnosis of malformation and volvulus. The UGI will show a misplaced duodenum with the ligament of Treitz on the right side of the abdomen, a duodenum with a "corkscrew" appearance, and duodenal obstruction, which may appear similar to that seen with duodenal atresia or may present with a "beak" appearance if a volvulus is present. An UGI is the gold standard for imaging to diagnose malrotation.

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Historically, a barium enema was considered the procedure of choice in these circumstances, but it has several limitations. Most importantly, the cecum can be in the proper position in the patient with duodenojejunal malrotation, and the latter can be missed. As the risk for midgut is related to duodenojejunal malrotation and not jejunoileal, the barium enema cannot be depended on to reach the affected portion of the intestines.

If the medical center has an experienced pediatric radiologist available, an **abdominal ultrasound** may be performed as the initial imaging. The findings on newborn ultrasound suggestive of malrotation include: a) the 3rd part of the duodenum is not in the normal retro-mesenteric position (between the mesenteric artery and the aorta in the retroperitoneal space); b) abnormal position of the superior mesenteric vein; and c) the "whirlpool" sign of volvulus caused by the vessels twisting around the base of the mesenteric pedicle, dilated duodenum indicating duodenal obstruction by Ladd's bands, duodenal obstruction with distal air. Magnetic resonance imaging (MRI) is more reliable for confirming intestinal malrotation than computed tomography (CT) and avoids ionizing radiation (e.g., pregnant patients). However, experience with this modality is limited, and practical matters such as sedation and length of study limit its usefulness in clinical situations where the diagnosis of volvulus is being entertained. Laparoscopy is a less invasive option than laparotomy for determining the presence of volvulus in equivocal situations.

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Suggested Reading:

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