1. A 12-day-old male premature infant presents with increasing bilious gastric aspirates and abdominal distension. You are shown an AP radiograph of the abdomen. What is the MOST likely diagnosis?

A. Necrotizing enterocolitis
B. Malrotation with volvulus
C. Hirschsprung’s disease
D. Meconium ileus

Findings:
Multiple bowel loops are dilated with gas, distending the abdomen. Several bowel loops in the lower abdomen show curvilinear lucency at their margins consistent with intramural gas (pneumatosis intestinalis). Bowel in the right lower quadrant has a “bubbly” appearance, also consistent with pneumatosis intestinalis. No free intraperitoneal air is evident.

Rationales:
A. Correct. In a neonate, this appearance is most consistent with necrotizing enterocolitis (NEC). The etiology of NEC is multifactorial. Hypoperfusion and stasis contribute to bacterial overgrowth and necrosis. The radiographic hallmark of NEC is intramural gas; however, many infants with NEC do not demonstrate this finding. Intramural gas may lead to portal venous gas. Perforation may produce pneumoperitoneum. Diffuse or focal gaseous distension of bowel is a common finding, but is less specific. The differential diagnosis for intramural gas in a neonate is very limited. The finding is highly suggestive, if not diagnostic, for NEC.
B. Incorrect. Although the clinical presentation of bilious gastric aspirates might suggest malrotation, usually these infants do not present with abdominal distention. Children with malrotation and malrotation with volvulus often have a normal bowel gas pattern or an evidence of a proximal obstruction. Diffuse gaseous distension of bowel, as in this child, from malrotation
is exceptionally rare. It has been reported that with gut necrosis from volvulus resorption of luminal gas ceases and gaseous distension results. Such children will be severely ill. Intramural gas from malrotation is also exceptionally rare, but can occur due to ischemic gut from volvulus. The bowel gas pattern in this child is therefore very atypical of malrotation or malrotation with volvulus. Moreover, when this pattern does occur with malrotation with volvulus, the infant will be severely ill, which is not the history provided.

C. Incorrect. Hirschsprung’s disease may produce a distal bowel obstruction leading to diffuse dilatation. Most of these children will present in the first few days of life with distension or failure to pass meconium, but presentation at 12 days of life is possible. Intramural gas, especially to this extent would be very unusual for Hirschsprung’s disease. Although published differential diagnosis lists for intramural gas often list obstruction as a cause, it is very rare for congenital bowel obstructions, including Hirschsprung’s disease, to produce intramural gas, especially to the degree seen in this child.

D. Incorrect. Meconium ileus is a presenting manifestation in approximately 10% of patients with cystic fibrosis. Tenacious meconium obstructs the distal ileum. The distal obstruction may cause diffuse upstream bowel dilatation; however, as discussed above, intramural gas would be very unusual. Moreover, careful inspection of the air-filled bowel loops in this patient does suggest that some are colon, and hence distal to the ileum mitigating against a distal ileal obstruction. The “bubbly” appearance of intramural gas in the right lower abdomen appears similar to the mottled appearance produced by gas mixed with meconium in a child with meconium ileus; however, this does not account for the gas seen with a curvilinear appearance elsewhere.

Citations:

2. Regarding congenital anomalies of the spinal cord, which of the following is CORRECT?

A. There is an increased incidence of tethering in patients with anal atresia.
B. Imaging of the cord is needed in infants with a low-lying coccygeal dimple.
C. Lipomyelomeningocele is associated with Arnold-Chiari malformation.
D. Skin-covered lesions are not associated with tethering.
Rationales:
A. Correct. Patients with anal atresia are at high risk of occult cord anomalies, resulting in tethering, and should undergo screening.
B. Incorrect. Low-lying dimples, unassociated with skin tags, skin discoloration, or hair tufts, are at low risk of tethering, and routine screening for these patients is not recommended.
C. Incorrect. Unlike the open defects, such as meningomyelocele, patients with lipomyelomeningocele do not typically have associated Chiari malformation.
D. Incorrect. Skin-covered cord lesions, including the lipomyelocele and lipomyelomeningocele, result in cord tethering and symptoms resulting from cord ischemia due to stretching of the cord.

Citations:

3. A neonate presents with complex congenital heart disease. Bilateral minor fissures are evident on chest radiographs. Which one of the following associated findings is MOST likely?
A. Biliary atresia
B. Malrotation
C. Interrupted inferior vena cava
D. Multiple spleens

Rationales:
A. Incorrect. Biliary atresia is seen in patients with polysplenia. This child has a heart lesion and trilobed lungs suggesting asplenia. Even in polysplenia, only a small fraction of children are affected. Thus, biliary atresia is not an expected abdominal finding in this child.
B. Correct. Although congenital heart disease is seen both in polysplenia and asplenia, the complex lesion in this child is more inconsistent with the severity of defects typically seen with asplenia than polysplenia. Children with polysplenia more often have simple shunts or anomalies of systemic or pulmonary return whereas those with asplenia often have transposition, common ventricles, and common atria. Bilateral minor fissures are indicative of bilateral trilobed lungs, which are consistent with asplenia syndrome (bilateral right-sidedness). Bilateral bilobed lungs are consistent with polysplenia syndrome (bilateral leftsidedness); however, often the fissures are difficult to see on neonatal films and thus lack of visualization of the minor fissure(s) does not equate to a diagnosis of polysplenia syndrome. Malrotation is present in most children with asplenia and thus would be an expected abdominal finding in this
child. It should be noted that malrotation is also present in most children with polysplenia and thus is not a differentiating feature.

C. Incorrect. Interrupted inferior vena cava is a feature of polysplenia syndrome. This child has a heart lesion and trilobed lungs suggesting asplenia. In asplenia syndrome, the inferior vena cava is uninterrupted, although it is often malpositioned to the left rather than the normal location to the right of midline. Thus, an interrupted inferior vena cava is not an expected abdominal finding in this child.

D. Incorrect. Multiple spleens, of course, are consistent with polysplenia, not asplenia. Often the spleens are found in the right upper quadrant rather than the left, so absence of the spleen in its normal location is not sufficient evidence of asplenia. This child has a heart lesion and trilobed lungs suggesting asplenia, not polysplenia. Thus, multiple spleens are not an expected abdominal finding in this child.

Citations:

4. Regarding head sonography in the premature infant, which one of the following is CORRECT?

A. Periventricular leukomalacia manifests as cystic lesions.
B. Grade I germinal matrix hemorrhage is found at the occipital horns.
C. Lack of sulcation is suggestive of lissencephaly.
D. A cavum septum pellucidum suggests lobar holoprosencephaly.

Rationales:
A. Correct. Periventricular leukomalacia is an ischemic/hemorrhagic injury in the periventricular white matter, which evolves into multiple cystic spaces visible on sonography.
B. Incorrect. Grade I bleed is confined to the germinal matrix, and is typically found anterior to the foramina of Monro. By definition, blood does not extend into the ventricles.
C. Incorrect. Premature infants typically lack sulcation of the brain.
D. Incorrect. Cavum septum pellucidum and cavum vergae are normal findings at sonography in the premature infant.

Citations:
5. Regarding pyloric stenosis, which one is TRUE?

A. It is present at birth  
B. Affected infants present with paradoxical aciduria  
C. Paternal history is a greater risk factor than maternal history  
D. Affected infants have no appetite

Rationales:
A. Incorrect. Pyloric stenosis develops after birth, and typically presents in infants at 4-12 weeks of age.
B. Correct. Vomiting leads to dehydration and waste of sodium, potassium and hydrochloric acid. Renal mechanisms supervene, with conservation of sodium at the expense of hydrogen ion, leading to a paradoxical aciduria in a patient with hypochloremic alkalosis.
C. Incorrect. Male and female children of affected mothers carry a 20% and 7% respective risk of developing the condition, whereas male and female children of affected fathers carry a respective risk of 5% and 2.5%.
D. Incorrect. Despite the distended stomach, the infants are malnourished, and exhibit a voracious appetite, typically gnawing at their hands in an effort to obtain nourishment.

6. You are shown a sagittal sonographic image of the pelvis of a 12-year-old girl with intermittent pelvic pain. What is the MOST LIKELY diagnosis?

A. Rhabdomyosarcoma  
B. Tuboovarian abscess  
C. Hematometrocolpos  
D. Hemorrhagic ovarian cyst
RATIONALES:
A. Incorrect. Cystic mass with debris levels as well as age of presentation are not consistent with typical findings of sarcoma botryoides. Multicystic vaginal tumor most typically presents between 1-3 years of age.
B. Incorrect. Cystic mass is posterior to the bladder – this location is more consistent with a vaginal or uterine origin rather than ovarian or fallopian. Cystic mass has the typical appearance of a dilated vagina with a fluid/fluid level, capped by the uterus with a dilated cervical canal and lumen.
C. Correct. The dilated fluid / debris filled structure posterior to the bladder is most consistent with a hematometrocolpos in the adolescent.
D. Incorrect. Cystic mass is posterior to the bladder and low in the pelvis. The location is more consistent with vaginal or uterine origin rather than ovarian. Further, an ovarian cyst could not have the typical appearance of a dilated vagina capped by a dilated uterine cervix and body.

References:

7. Which of the following is TRUE concerning sacrococcygeal teratomas?
A. Excision of the lower sacrum is necessary to prevent recurrence.
B. Currarino triad includes sacrococcygeal teratoma, sacral defect and posterior meningoloecele.
C. Sacrococcygeal teratomas contain elements of all three germ cell layers.
D. A type IV lesion is located below the sacrum and is visible externally.

RATIONALES:
A. Incorrect. Excision of the coccyx is necessary. Incomplete excision of the coccyx can result in recurrence in up to 37% of cases.
B. Incorrect. Associated with anterior meningomyeloceles
C. Correct. The hallmark of this tumor is that it contains elements from endoderm, mesoderm and ectoderm.
D. Incorrect. Type I lesion is external; Type II lesion contains an intrapelvic component; Type III is predominantly intrapelvic with an external component; Type IV lesion is located entirely within the pelvis.

References:
8. Which of the following represents an absolute CONTRAINDICATION to attempted image-guided reduction of intussusception in a 2-year-old child?

A. Pathological lead point
B. Ascites
C. Peritoneal signs
D. History of multiple prior reductions

Rationales:
A. Incorrect. A lead point may make the reduction statistically less likely to be successful, but a successful reduction would make surgery for the lead point elective.
B. Incorrect. Simple free fluid is often noted on ultrasound and does not correlate with a risk of perforation.
C. Correct. Peritoneal signs are indicative of peritonitis, consistent with bowel necrosis and perforation. Surgery is indicated without preceding attempt at reduction.
D. Incorrect. Some children have recurrent idiopathic intussusceptions. Recurrence should prompt consideration of careful search for a pathologic lead point, but does not exclude attempted reduction acutely.
9. You are shown a sagittal image of a spinal US over the lumbosacral region of a 2 week old infant. What is the diagnosis?

A. Ventriculus terminalis
B. Terminal myelocystocele
C. Lipomyelocele
D. Lipomyelomeningocele

Rationales:
A. Incorrect. Ventriculus terminalis refers to a normal variant, consisting of a small, oval, cystic structure located at the tip of a normally-positioned conus. This is not present in this case, and this choice is incorrect.
B. Incorrect. Terminal myelocystocele is a rare form of spinal dysraphia, often associated with caudal regression syndromes. It consists of dilatation of the terminal portion of the central canal of a tethered cord, with dilatation of the adjacent subarachnoid spaces. These findings are not present in this case, and therefore this choice is incorrect.
C. Correct. The findings are those of a tethered cord, with encroachment of subcutaneous fat through the spina bifida defect into the spinal canal and neural placode
D. Incorrect. In patients with lipomyelomeningocele, there is dilatation of the subarachnoid space, elevating the neural placode. These findings are not present in this case. In this characteristic, lipomyelocele and lipomingomyelocele are analogous to myelocoeles and myelomenigoceles, respectively.
10. Which lesion is commonly a vascular ring?

A. Pulmonary sling
B. Left aortic arch with aberrant right subclavian artery
C. Right aortic arch with aberrant left subclavian artery
D. Right aortic arch, mirror image branching

Rationales:
A. Incorrect. With a pulmonary sling, the left pulmonary artery is anomalous, originating from the right pulmonary artery and then passing posterior to the trachea and anterior to the esophagus to reach the left hilum. This does not constitute a vascular ring. Children with pulmonary sling do often have associated deformity of the trachea and proximal bronchi including anomalies, stenosis, complete cartilage rings and tracheobronchomalacia. These airway abnormalities may result in respiratory symptoms.
B. Incorrect. Left aortic arch with aberrant right subclavian artery is usually an asymptomatic normal variant in children. The ligamentum arteriosum is on the left. Therefore, there is no “ring” encircling the trachea. The aberrant right subclavian artery causes a posterior impression on the esophagus.
C. Correct. With a right aortic arch and an aberrant left subclavian artery, the ligamentum arteriosum (on the left) completes the vascular “ring” encircling the trachea and esophagus. The degree of symptoms depends on how tight the ring is and the degree of tracheal compression caused by the ring. Infants and young children usually present with airway symptoms related to vascular ring rather than symptoms related to esophageal compression.
D. Incorrect. With mirror image right aortic arch, aortic branching mirrors that of the left aortic arch with the (left) brachiocephalic artery originating first, followed by right carotid artery and right subclavian artery. Since the left subclavian artery originates from the brachiocephalic artery, there is no vessel passing posterior to the trachea and esophagus to complete a vascular ring.

11. Which of the following is an absolute CONTRAINDICATION to attempting enema reduction of an intussusception?

A. A 3-month-old patient
B. Symptoms for 24 hours
C. Intussusception to the rectum
D. Peritoneal signs
Rationales:
A. Incorrect. Presentation with intussusception at three months of age is atypical, and at the lower end of the age spectrum. The younger the patient, the greater the likelihood of an underlying lead point. Nevertheless, the age of the patient is not a contraindication to attempting reduction of the intussusception with an enema.

B. Incorrect. Extended symptoms indicate that the intussusception has probably been present for a longer duration. This indicates a lesser likelihood of successful reduction with enema. It is not an absolute contraindication to attempting reduction of the intussusception with an enema.

C. Incorrect. Ileocolic intussusceptions may extend all the way through the colon and even prolapse out of the patient. An intussusception extending to the rectum is less likely to be successfully reduced than one in the proximal colon. There is also probably an increased incidence of perforation with such cases. Nevertheless, an intussusception extending to the rectum is not an absolute contraindication to attempting reduction of the intussusception with an enema. Even if the intussusception is only partially reduced, this makes the surgical management easier.

D. Correct. The presence of peritoneal signs indicates a high likelihood of ischemia of the intussuscepted bowel and/or perforation. In such patients, successful reduction without complication (perforation) is unlikely and surgical management is indicated.