28th Annual

In-Training Examination for Diagnostic Radiology Residents Rationales

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Section X – Pediatric Radiology

Figure 1A

Figure 1B
260. You are shown two images (Figures 1A and 1B) from a contrast-enhanced abdominal CT in an 8-year-old child. What is the MOST likely diagnosis?

A. Pyelonephritis
B. Autosomal recessive polycystic kidney disease
C. Leukemia
D. Nephroblastomatosis

Question #260

Rationales:

A. Incorrect. Although pyelonephritis would present as poorly opacified renal segments on CT, these areas would not spare the cortex, as in the test case. Pyelonephritis typically causes perinephric inflammation, with stranding of the perinephric fat, which is absent here. Furthermore, such a diagnosis would not explain the splenomegaly, or the varices seen on the accompanying image.

B. Correct. Autosomal recessive polycystic kidney disease consists of abnormal tubules within the kidney, as in the test case, where the abnormality is seen to be located in the renal medulla. In an 8-year-old child, changes of hepatic fibrosis, with varices and splenomegaly, are characteristic.

C. Incorrect. Although leukemia can present with splenomegaly and renal involvement, leukemia does not lead to varices, or to the characteristic renal medullary changes seen here.

D. Incorrect. Nephroblastomatosis consists of multiple masses within the kidneys. The renal abnormality on the CT does not consist of masses; splenomegaly and varices are not part of this entity.

Citations:
261. You are shown an image of the bladder (Figure 2A) from a postnatal ultrasound, and two images (Figures 2B and 2C) from a VCUG on a male neonate with prenatal diagnosis of hydrenephrosis. What is the MOST likely diagnosis?

A. Hutch diverticulum
B. Posterior urethral valves
C. Evertting ureterocele
D. Primary obstructing megaureter
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**Question #261**

**Rationales:**

A. *Incorrect.* Although Hutch diverticulum protrudes from the bladder as a contrast-filled structure on VCUG, it does not appear as solid tissue within a fluid-filled structure separate from the bladder on Sonography, nor as a filling defect in the bladder, as in Fig. 2B

B. *Incorrect.* Posterior urethral valves are diagnosed during voiding, and these are not voiding images. The bladder is smooth-walled, and its wall is not thickened, as would be expected with posterior urethral valves. Posterior urethral valves also would not explain the smooth filling defect within the bladder seen in Figure 2B, which is characteristic of ureteroceles.

C. **Correct.** Evert fold ureterocele is a condition seen when the ureterocele, which is the dilated intramural portion of the ureter, everts back into the extravesical ureter, outside of the bladder lumen. The sonogram shows the dilated, fluid-filled ureter, containing the everted and intravesical portion. The fluoroscopic image shows the smooth filling defect within the bladder initially (Fig. 2B) and everted on the subsequent image (2C) with the contrast-filled ureterocele protruding outside of the bladder.

D. *Incorrect.* Primary obstructing megaureter refers to obstruction of the distal ureter due to a short juxtavesical aperistaltic segment. Such a segment contains only anechoic urine on sonography, and does not have a blunted end, if reflux were to be concurrently present. Further, such a diagnosis also would not explain the smooth filling defect within the bladder, seen in Figure 2B.

**Citations:**


262. 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 (Figure 3). What is the MOST likely diagnosis?

A. Necrotizing enterocolitis
B. Malrotation with volvulus
C. Hirschsprung’s disease
D. Meconium ileus
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Question #262

Findings:
Multiple bowel loops are dilated with gas, distending the abdomen. Several bowel loops in the left 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. Stasis and hypoperfusion contribute to bacterial overgrowth and necrosis.

The radiographic hallmark of NEC, although not visible in all infants, is intramural gas. 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, and the infant presents soon after birth. The distal obstruction may cause diffuse upstream bowel dilatation; however, as discussed above, intramural gas would be very unusual. Moreover, careful inspection of the radiograph shows stool in the rectum, and hence distal to the ileum excluding 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:

263. You are shown an extremity radiograph (Figure 4) on a 4-month-old with irritability and altered mental status. What is the MOST likely diagnosis?

A. Rickets
B. Congenital syphilis
C. Non-accidental trauma
D. Scurvy
Findings:
The findings consist of periosteal reaction around the metaphyses of the distal femur and the proximal tibia. The periosteal reaction around the metaphyses is circumferential, indicative of metaphyseal linear injury about the metaphyseal spongiosa. This results in the appearance of “bucket handle” density about the metaphyses, typical of nonaccidental trauma or battered child syndrome. There is exaggeration of periosteal reaction about the adjacent diaphyses, suggesting repetitive injury and calcified hemorrhage.

Rationales:
A. Incorrect. Rickets is the result of deficiency of vitamin D, and in turn leads to failure of ossification of the physeal cartilage. This leads to loss of the radiodense zone of provisional calcification, and fraying of the metaphyses. Failure of mineralization leads to a widened appearance of the physis. These features are lacking in the test case.

B. Incorrect. Congenital syphilis is a transplacently acquired infection, during the second or third trimester of pregnancy.

A negative maternal syphilis test early in gestation therefore may not exclude the diagnosis in the infant, since maternal infection could have been acquired later in pregnancy. Skeletal manifestations typically develop in the infant at 6 – 8 weeks of age. Although syphilis may manifest s diffuse periostitis with periosteal reaction, the typical findings that are also present include radiolucent metaphyses, and destructive lesions in the medial tibial metaphysis, known as Wimberger sign. These typical findings of congenital syphilis are not present in the test case.

C. Correct. Correct answer as listed in the findings above.

D. Incorrect. Vitamin C deficiency, or scurvy, results in inability to synthesize normal collagen. The appearance of the bones is characterized by osteoporosis, with thin, sharp cortices. In the epiphyses, the outer border is sharply marginated, resembling a thin ring, the Ring of Wimberger. The zones of provisional calcification are preserved, but are underlined by a subjacent zone of mineralization, termed the scurvy line. Fragility of the tissues can result in extensive subperiosteal bleeding, and this can exhibit exuberant periosteal calcification during the healing phase. Although the periosteal reaction in the test case can resemble the calcification of subperiosteal hemorrhage in scurvy, this does not occur until the healing phase. Further, other findings of vitamin C deficiency—the lucent zone, Wimberger’s Ring, and diffuse osteoporosis—are absent.
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Figure 5A

Figure 5B

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264. You are shown coronal ultrasound (Figure 5A) and T1-weighted axial MR (Figure 5B) images of the brain in a six-month-old with enlarging head. What is the MOST likely diagnosis?

A. Posterior fossa hemorrhage
B. Ependymoma
C. Cystic astrocytoma
D. Vein of Galen aneurysm

Question #264

Findings:
Sonogram demonstrates a hypoechogenic mass in the posterior fossa with associated hydrocephalus. MRI demonstrates a signal void in the posterior fossa with associated pulsatile artifacts confirming the presence of a vascular lesion.

Rationales:
A. Incorrect. While rare, posterior fossa hemorrhages can occur in preterm and term infants following hypoxic ischemic injury. Posterior fossa hemorrhages have been particularly associated with term infants on extracorporeal membrane oxygenation. While the ultrasound image could suggest a hematoma within the posterior fossa, the low signal on the T1-weighted image is consistent with flowing rather than clotted blood, and signs of pulsatility. Doppler images by ultrasound can help document turbulent flow in the hypoechogenic posterior fossa mass confirming the diagnosis of an arteriovenous malformation.

B. Incorrect. Ependymomas are neoplasms of the ciliated ependymal cells arising from the ependymal lining of the ventricles. They are relatively slow growing, infiltrating, and difficult to remove surgically. Most commonly they are infratentorial situated in the floor of the fourth ventricle. By CT a mass is often noted in the region of the fourth ventricle associated with obstructive hydrocephalus. Calcifications are frequent. Unlike the test case, ependymomas are typically inhomogeneous with variable hypointensity on T1W.

C. Incorrect. Cerebellar astrocytomas are derived from astrocytic neuroglial cells and tend to be cystic and well circumscribed. On CT images, a well-defined area of fluid with an isodense mural nodule is typically noted. The cyst contents are more intense that CSF due to the high protein and thus are of higher signal on T1W than the adjacent CSF. The mural nodule and rim of the cyst will enhance intensely following gadolinium administration.

D. Correct. Vein of Galen aneurysms are actually arteriovenous malformations, with “aneurysmal” dilatation of the vein of Galen, rather than a true aneurysm. There are usually aberrant connections between choroidal arteries thalamoperforators and vein of Galen. Infants may present with congestive heart failure due to high output failure. Doppler studies can confirm the presence of a high-flow vascular mass. MRI demonstrates low signal due to flow void. Contrast will demonstrate intense enhancement of the aneurysm. Treatment can be successful via interventional embolization

Citations:
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265. Regarding congenital anomalies of the spinal cord, which one 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.

Question #265

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 necessary.
C. Incorrect. Unlike the open defects, such as meningomyelocele, patients with lipomyelomenigocele do not
   typically have associated Chiari malformation.
D. Incorrect. Skin-covered cord lesions, including the lipomyelocele and lipomyelomenigocele, result in cord
   tethering and symptoms resulting from cord ischemia due to stretching of the cord.

Citations:
266. Concerning vascular rings, which one of the following is CORRECT?

A. Double aortic arch usually presents during puberty.
B. The double aortic arch encircles the trachea anterior to the esophagus.
C. Symptomatic pulmonary sling is associated with a right aortic arch.
D. Left aortic arch with aberrant right subclavian artery is typically asymptomatic.

**Question #266**

**Rationales:**

A. *Incorrect.* Double aortic arch is a true vascular ring resulting in compression of both trachea and esophagus. Symptoms often begin at birth and can include stridor, wheezing, vomiting, and dysphagia.

B. *Incorrect.* Double aortic arch typically encircles both the trachea anteriorly and esophagus posteriorly by its right and left arches. Lateral radiographs will demonstrate anterior bowing of the trachea. On barium swallow posterior indentation of the esophagus is present.

C. *Incorrect.* Anomalous origin of the left pulmonary artery is often an isolated finding though it may be part of a more complex anomaly, with tracheal stenosis. The left pulmonary artery arises from the right pulmonary artery crossing over the proximal portion of the right mainstem bronchus.

D. *Correct.* This most common congenital vascular anomaly is frequently found due to interruption of the right embryonic arch between the right common carotid and right subclavian arteries. The right ductus arteriosus usually involutes so no true vascular ring exists.

**Citations:**


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267. 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

Question #267

Rationales:
A. *Incorrect.* This child has complex congenital heart disease and trilobed lungs. This indicates situs ambiguous, or heterotaxy. Further, the bilateral trilobed lungs are very strongly suggestive of bilateral right-sided complex, or asplenia syndrome. Biliary atresia is seen in some patients with polysplenia, but not in asplenia. Thus, biliary atresia is not an expected abdominal finding in this child.

B. *Correct.* Malrotation is seen in children with asplenia and with polysplenia, and thus would be an expected abdominal finding in this child.

C. *Incorrect.* Interrupted inferior vena cava is a feature of polysplenia syndrome. This child has 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. The splenules are located along the greater curvature of the stomach. This child has trilobed lungs suggesting asplenia, not polysplenia. Thus, multiple spleens are not an expected abdominal finding in this child.

Citations:


268. 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.

Question #268

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 in Grade I bleeds.
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. In holoprosencephaly, the septum pellucidum is absent.

Citations:
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269. Concerning developmental hip dysplasia, ALL of the following are true EXCEPT:
   A. Female infants are more commonly affected.
   B. It is more common with breech deliveries.
   C. Hip sonography screening is most useful in the first week of life.
   D. Normal alpha angle is greater than 60 degrees.

Question #269

Rationales:
A. Incorrect. The choice is True. Female infants are at higher risk for hip instability with a ratio of about 8:1.
B. Incorrect. The choice is True. Risk of hip dysplasia in breech deliveries is 6 times higher than vertex births.
C. Correct. The choice is False. Normal neonatal hip laxity may lead to inaccurate results. Laxity in the early days after birth may be a hormonal effect produced by maternal estrogens that are not completely inactivated by the immature fetal liver. Thus, hip ultrasound screening is recommended by 4-6 weeks of age.
D. Incorrect. The choice is True.

Citations: