132. A 1-day-old presents with abdominal distention. Based on the image (Figure 1), what is the MOST LIKELY diagnosis?

A. Hirschsprung’s disease
B. Meconium ileus
C. Ileal atresia
D. Meconium plug syndrome

Findings:
Contrast enema demonstrates a microcolon with contrast material refluxing into the terminal ileum which is filled with multiple filling defects consistent with pellets of meconium. Findings are most consistent with meconium ileus.

Rationale:
A: Hirschsprung disease typically presents with a normal sized colon and may have a visible transition zone, often at the rectosigmoid. Total colonic anganglionosis is rare, and while it can rarely present with microcolon, should not have meconium filling the distal ileum.
B: There is a microcolon with contrast refluxing into slightly dilated terminal ileum filled with meconium pellets. This helps separate this diagnosis from ileal atresia and total aganglionosis. This diagnosis is almost always associated with cystic fibrosis.
C: While ileal atresia will present with microcolon, contrast should not reflux into dilated meconium filled loops of ileum.
D: Meconium plug syndrome should have normal sized colon filled with long meconium plug.
133. Concerning the finding shown on the CT image (Figure 2) of a 1-year-old, which of the following statements is TRUE?

A. Calcifications are present in 30% of cases.
B. The hereditary form is unilateral and unifocal.
C. The majority of cases are autosomal dominant.
D. The trilateral form is associated with neuroectodermal pineal tumors.

**Findings:**
CT demonstrates a calcified mass within the left orbit. Intraocular calcification in children less than 3 years of age is highly suggestive for retinoblastoma. Other etiologies include CMV chorioretinitis, Colobomatous cyst. In children older that 3 years of age differential includes toxocariasis, drusen of the optic nerve head and retinal astrocytoma.

**Rationale:**
A: Calcifications are present in over 90% of cases.
B: While the nonhereditary form is unilateral, and unifocal, the hereditary form in typically bilateral and multifocal.
C: Autosomal dominant risk factors linked to chromosome 13 are found in 10% of affected children. Numerous tumors, including melanoma and fibrosarcoma, have been associated with children with a genetic tendency to retinoblastoma.
D: Trilateral retinoblastoma typically present with bilateral ocular retinoblastomas and a neuroectodermal tumor in the pineal such as a pinealoblastoma.
134. Based on the sagittal MR image (Figure 3), the tumor depicted in the fetus would be considered:

A. Type I.
B. Type II.
C. Type III.
D. Type IV.

Findings:
Lobular cystic mass between the sacrum and rectum, with extrapelvic and intrapelvic components, the latter confined to the pelvis.

Rationale:
A: Sacrococcygeal teratoma can be primarily cystic, as in this case, primarily solid, or mixed. Typing is based on a surgical approach to the lesion. Type I SCT is defined as essentially extrapelvic, with very little, if any, presacral component.
B: Type II lesions are those with both extra- and intrapelvic components. The pelvic tumor should not extend into the abdomen, as removal of any significant abdominal component would dictate a surgical approach different from that required to excise a tumor confined to the pelvis.
C: Type III lesions are primarily intrapelvic and tumor involvement extends from the pelvis into the abdomen.
D: These lesions historically have been detected later than the others because they are entirely intrapelvic, with no external component at all. This accounts for their higher rate of malignancy when (eventually) found.
135. A newborn presents with seizures. Based on the image (Figure 4), what is the MOST LIKELY diagnosis?

A. Inferior vermian hypoplasia  
B. Gray matter heterotopia  
C. Callosal agenesis  
D. Cavum velum interpositum

**Findings:**
Radiating gyri in a sunburst pattern, and no corpus callosum on the midline sagittal image.

**Rationale:**
A: In this midline sagittal view, the echogenic vermis for the cerebellum appears fully formed caudally, practically touching the brainstem. The fourth ventricle appears normal.
B: On ultrasonography, heterotopia might be detected by noting a nodular appearance to the margins of the lateral ventricles. This diagnosis would not be made from a sagittal midline image.
C: This image demonstrates a characteristic appearance of outward radiation of medial hemispheric gyri, pointing toward the third ventricle, without a corpus callosum or cingulate gyrus.
D: A CVI is an expansion of the velum interpositum cistern, and would be seen on ultrasound as a fluid collection between the region of the pineal and the dorsal corpus callosum. It is a normal variant and would be detected on a midline sagittal image, but is not present in this case.
136. A 21-month-old girl presents with fever and malaise (Figures 5-7). What is the MOST LIKELY diagnosis?

A. Retroperitoneal teratoma  
B. Wilms’ tumor  
C. Neuroblastoma  
D. Desmoplastic small round cell tumor

Findings:  
The images show a very large, solid retroperitoneal mass which depresses the ipsilateral kidney, and encases and displaces the aorta and its branches.

Rationale:  
A: Although a retroperitoneal teratoma would occur in this location, it would be expected to contain various tissue types, including cystic areas, calcifications, and areas of fat. In addition, it is much less common than neuroblastoma.  
B: Although Wilms tumor does occur in this age group, the images show that this mass is not intrarenal, but instead arises above and displaces the kidney.  
C: The images show a very large, solid retroperitoneal mass which depresses the ipsilateral kidney, and encases and displaces the aorta and its branches. These findings, particularly in this age group, are typical of neuroblastoma.  
D: Although this lesion can occur in the retroperitoneum, it is rare, much more common in boys (approximately 96%), and typically occurs in adolescents; it presents more commonly in the pelvis.
137. A 7-year-old boy has a limp. Based on the radiographs (Figures 8 and 9), what is the MOST LIKELY diagnosis?

A. Developmental hip dysplasia  
B. Leukemia  
C. **Legg-Perthes disease**  
D. Child abuse

**Findings:**  
The images demonstrate increased density and loss of height of the left femoral head. In addition, on the frog lateral view, a crescent-shaped subchondral lucent line is visible, which represents a subchondral fracture of weakened, dead bone.

**Rationale:**  
A: In developmental hip dysplasia, the acetabulum is shallow or non-existent, and the femoral head is subluxed or dislocated, articulating with a pseudoacetabulum in the ilium.  
B: Although patients with leukemia can present with bone pain, at times localized to the hip, in patients with leukemia, there is overall osteopenia, which is not present here, and there are radiolucent metaphyseal lines, often termed "leukemic" lines, which are also not present here.  
C: The images demonstrate increased density and loss of height of the left femoral head. In addition, on the frog lateral view, a crescent-shaped subchondral lucent line is visible, which represents a subchondral fracture of weakened, dead bone. These findings are typical of Legg-Perthes disease in this age group.  
D: In abused patients, skeletal findings include fractures of different ages, and rib and corner fractures, typically in younger patients.
138. A CT scan (Figures 10 and 11) is performed on a 12-month-old girl with a palpable right lower quadrant mass. Which of the following statements is TRUE of the entity shown?

A. There are two orthotopic ureteral orifices.
B. The left ureteral orifice is ectopic.
C. The right ureteral orifice is ectopic.
D. The bladder has only one ureteral orifice.

Findings:
The images show fusion of the left kidney to the lower pole of the right kidney.

Rationale:
A: The images show fusion of the left kidney to the lower pole of the right kidney. These are findings of crossed-fused renal ectopia. In such cases, each kidney drains through its own orthotopic ureteral orifice.
B: Incorrect.
C: Incorrect.
D: Incorrect.
139. A coronary CTA (Figure 12) is performed on a 7-year-old boy. What is the MOST LIKELY diagnosis?

A. Aberrant right coronary  
B. Aberrant left coronary  
C. Kawasaki’s disease  
D. Marfan’s syndrome

**Findings:**
The image is an oblique reconstruction of the course of the right coronary artery, and the proximal portion of the left coronary artery. Both of these arteries show prominent aneurysmal dilatations; there is no contrast within a portion of the course of the right coronary, indicating that there has been thrombosis of another aneurysm.

**Rationale:**
A: The right coronary clearly arises from the right coronary cusp  
B: The left coronary clearly arises from the left coronary cusp  
C: The image is an oblique reconstruction of the course of the right coronary artery, and the proximal portion of the left coronary artery. Both of these arteries show prominent aneurysmal dilatations; there is no contrast within a portion of the course of the right coronary, indicating that there has been thrombosis of another aneurysm. Coronary artery aneurysms are one of the well-known complications of Kawasaki disease.  
D: Although Marfan leads to dilatation of the aortic root, coronary artery aneurysms are very rare, and reportedly described only in adults.
140. A 4-week-old infant presents with febrile UTI. Based on the images (Figures 13 and 14), what is the MOST LIKELY diagnosis?

A. Duplex system on the left
B. Duplex system on the right
C. Bilateral duplex systems
D. Bilateral single systems

Findings:
The voiding image shows a left-sided ectopic ureter arising from the urethra, which fills during voiding. The post void image shows reflux into the left sided upper pole drained by the ectopic ureter. It also shows reflux into the right kidney, with "drooping lily" appearance.

Rationale:
A: Incorrect.
B: Incorrect.
C: The voiding image shows a left-sided ectopic ureter arising from the urethra, which fills during voiding. The post void image shows reflux into the left sided upper pole drained by the ectopic ureter. It also shows reflux into the right kidney, with "drooping lily" appearance. This appearance indicates that there is a duplex system on the left, with reflux into the left upper pole via the ectopic ureter, and also a duplex system on the right, with reflux only into the right lower pole via its orthotopic orifice.
D: Incorrect.
141. A chest radiograph (Figure 15) and sequential CT images (Figures 16-18) are shown of a neonate with an abnormal prenatal ultrasound. What is the MOST LIKELY diagnosis?

   A. Bronchial atresia
   B. Scimitar syndrome
   C. Airway foreign body with air trapping
   D. Left pulmonary artery sling

**Findings:**
Overinflated left upper lobe with mucous impaction in bronchus.

**Rationale:**
A: There is overinflation of the left upper lobe and mucous impaction in the bronchus, which can be seen and followed adjacent to the artery.
B: The affected lung is typically on the right and hypoplastic in scimitar syndrome, with abnormal venous drainage; mucous impaction would not be present.
C: An airway foreign body could cause lobar over-inflation and a soft tissue density in the airway, but this would not be seen in-utero as described in this case, nor would it be likely in a neonate or distally in the upper lobe bronchus.
D: A left pulmonary artery sling could cause air-trapping, but there is no evidence of an anomalous left pulmonary artery. Furthermore, bronchial mucoid impaction would not be seen.
142. Among the choices given below, which strategy has the MOST effect in reducing pediatric dose?

A. Using tube current modulation
B. Using low pitch values
C. Using a smooth reconstruction algorithm
D. Using higher tube voltage (kVp)

Rationale:
A: The use of tube current modulation allows the CT scanner to vary the tube current based on the patient thickness, scanning at lower tube current while scanning thinner part of the anatomy and higher tube current while scanning thick part of the anatomy and still maintaining consistent image quality.
B: Low pitch values increases radiation dose, since low pitch value has increased overlap of the anatomy
C: Reconstruction algorithms have no effect on the radiation dose in general
D: Increasing the tube voltage (kVp) will increase radiation dose to patient

143. Which of the following statements is TRUE concerning holoprosencephaly?

A. The septum pellucidum is absent in all forms of holoprosencephaly.
B. Interhemispheric fissure is absent in all forms of holoprosencephaly.
C. In the semilobar form, only the genu of the corpus callosum is present.
D. Cyclopia is associated with lobar holoprosencephaly.

Rationale:
A: There is no septum pellucidum in alobar, semilobar or lobar holoprosencephaly
B: The interhemispheric fissure may be partially present in semilobar and lobar holoprosencephaly.
C: Holoprosencephaly is the only congenital brain anomaly in which the posterior portion of the corpus callosum has formed in the absence of the anterior portion.
D: Facial anomalies are mild or absent with semilobar holoprosencephaly. Cyclopia is typically associated with the most severe form, or alobar holoprosencephaly.

144. Regarding neurofibromatosis type 1, which of the following statements is TRUE?

A. Three or more criteria are needed for diagnosis.
B. Long bones are not involved.
C. Axillary or groin freckling is frequent in younger patients.
D. A single neurofibroma is pathognomonic.

Rationale:
A: Two or more criteria are needed for diagnosis. Optic gliomas often develop in patients with NF1. Other CNS tumors that can develop include cerebellar and cerebral astrocytomas, brainstem glioma, and ependymomas.
B: Musculoskeletal manifestation are frequent in 50-70% of patients with NF1. They are a result of mesodermal dysplasia or direct erosive changes on bone by adjacent soft tissue tumors.
C: Freckling of the axilla or groin is essentially pathognomonic for NF1 and is found in 81% of patients younger than 6 years of age with NF1.
D: Neurofibromas and neurilemmomas are more common as isolated lesions not associated with NF1. However, when multiple and involving deep nerves they are associated with NF1.
145. Which one of the following ultrasound findings indicates developmental dysplasia of the hip (DDH)?

   A. Fifty-five percent coverage of the femoral head by bony acetabulum
   B. Graf alpha angle of 50°
   C. Anterior motion of femoral head on stress maneuver
   D. Very thin pulvinar

**Rationale:**
A: Femoral head coverage by the bony acetabulum in DDH is less than 50%; over 50% typically indicates a normal hip.
B: An alpha angle of less than 60 degrees indicates a steep acetabular roof and shallow acetabulum in DDH.
C: Posterior subluxation of the femoral head on the stress views may indicate instability and is associated with DDH. Anterior motion does not occur.
D: A hypertrophic, not a very thin, pulvinar may be seen in DDH, occupying space in the shallow acetabulum, rendering appropriate reduction more difficult.

146. Which of the following develops in progressive skeletal changes after the first year of life?

   A. Achondrogenesis
   B. Morquio’s syndrome
   C. Thanatophoric dysplasia
   D. Jeune’s syndrome

**Rationale:**
A: Achondrogenesis is a lethal skeletal dysplasia that has micromelia, short ribs, absent vertebral body ossification.
B: Morquio Syndrome (Mucopolysaccharidosis type IV) is the result of the accumulation of excess MPS material in multiple organ systems including the skeleton. Bony changes progress during the first few years of life.
C: Thanatophoric dysplasia is invariably lethal with severe micromelia, short ribs, and platyspondyly.
D: Asphyxiating Thoracic Dysplasia otherwise known as Jeune syndrome is a disorder with a mixed prognosis. Many affected patients die in the perinatal period due to their small chest. Survivors may die from renal complications later in life. The ribs are short and horizontal with short extremities and a normal spine.
147. Magnetic resonance imaging of the fetus relies MOST heavily on which of the following types of acquisitions?

A. Gradient echo
B. Single-shot fast/turbo-spin echo
C. Short tau inversion recovery (STIR)
D. Echo-planar imaging (EPI)

Rationale:
A: These can be used for T1 weighted images in select situations, however.
B: This type of sequence allows for sub-second image acquisition and the T2 weighting highlights pathology well.
C: Attempts at fat suppression imaging can be helpful in select situations but are not routine.
D: Occasionally used to emphasize bony structures.

148. Regarding congenital adrenal hyperplasia, which of the following statements is TRUE?

A. The most common form is 11-beta-hydroxylase deficiency.
B. There is elevation of serum cortisol levels.
C. It is inherited as an autosomal dominant trait.
D. Men can present with life-threatening salt-wasting crisis.

Rationale:
A: Approximately 90% of cases of congenital adrenal hyperplasia are secondary to a defect in the 21-hydroxylase enzyme.
B: Because of the blockage in the synthesis of cortisol due to the enzymatic deficiency, cortisol in these patients is diminished, with elevation of ACTH and adrenal hyperplasia.
C: Congenital adrenal hyperplasia is inherited as an autosomal recessive trait.
D: In the more severe forms of the enzyme deficiency, there is diminished production of aldosterone in addition to cortisol. Affected female infants present with masculinization of the external genitalia; however, in male infants initial presentation can be with a salt-wasting crisis, as they are not identified earlier by abnormal genitalia, as is the case in affected female infants.
149. What is the MOST LIKELY consequence of a conceptus radiation dose of 10 mSv during the major organogenesis period of gestation (2 to 8 weeks)?

A. Prenatal death  
B. Microcephaly  
C. Childhood cancer  
D. Congenital heart disease

**Rationale:**  
A: The threshold dose for prenatal death during this gestation period is higher than 10 mGy - perhaps > 250 mGy.  
B: Microcephaly in this gestation period has been observed for doses above 100 mGy.  
C: Childhood cancer risk is thought to have no lower threshold, though the risk is low – 6 in 10,000 for a 10 mSv exposure.  
D: The only organ system that has shown an association with radiation dose < 250 mGy is the central nervous system.

150. Which of the following requires surveillance imaging because of a high risk for the development of Wilms’ tumor?

A. Glomerulonephritis  
B. WAGR syndrome  
C. Horseshoe kidney  
D. Infants of mothers with diabetes mellitus

**Rationale:**  
A: Glomerulonephritis is not associated with increased risk of Wilms tumor development.  
B: WAGR syndrome, (Wilms tumor, aniridia, genitourinary anomalies and mental retardation) is associated with an 11p13 deletion encompassing the WT1 gene for Wilms tumor, and the PAX 6 gene for aniridia.  
C: Although horseshoe kidney may have a slightly increased incidence of Wilms tumor, it does not require surveillance imaging, as is the case in patients with sporadic aniridia, Beckwith-Wiedemann or nephroblastomatosis.  
D: Infants of diabetic mothers are not at risk for Wilms tumor.

151. Regarding renal scarring, which of the following statements is TRUE?

A. It does not occur in the absence of reflux.  
B. Older children are at greater risk.  
C. It is most common in boys.  
D. It can lead to hypertension.

**Rationale:**  
A: Infection and renal scarring can develop in 37-43% of children without demonstrable reflux.  
B: The younger the child, the greater the risk of scarring, particularly in patients younger than 4 years, and even more so when younger than 1 year.  
C: Renal infection and subsequent scarring is more common in girls.  
D: Chronic renal failure, and more commonly hypertension, can follow recurrent episodes of pyelonephritis and renal scarring.
152. Regarding bronchopulmonary sequestration, which of the following statements is TRUE?

A. The sequestered lung communicates with the tracheobronchial tree.
B. **Extralobar sequestrations are associated with multiple congenital anomalies.**
C. The majority of sequestrations are extralobar.
D. Intralobar sequestrations drain via the azygous system.

**Rationale:**
A: The involved segment of lung usually does not communicate with the tracheobronchial tree.
B: Extralobar sequestration may be associated with other lesions, such as diaphragmatic hernia, congenital heart disease, and CPAM (Cystic Pulmonary Airway Malformation)
C: Approximately 75% of cases are intralobar.
D: Intralobar sequestrations are usually drained by the pulmonary venous system.

153. Regarding Peutz-Jeghers syndrome, which of the following statements is TRUE?

A. The stomach is the most common site of polyps.
B. **There is an increased risk of malignancy.**
C. Most patients are symptomatic in the first decade.
D. Adenomatous polyps are the typical type of polyps.

**Rationale:**
A: The small bowel is the most common site of tumors in Peutz-Jeghers syndrome.
B: There is increased risk of GI malignancies as well as malignancies in the GU tract, breast, pancreas, thyroid, and skin (as well as multiple myeloma) in patients with Peutz-Jeghers syndrome.
C: Only one-third of patients with Peutz-Jeghers syndrome will experience symptoms during the first decade of life.
D: Hamaromatous polyps are the classic type of polyps in Peutz-Jeghers syndrome.