1. 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

**Rationale:**
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 a 4-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.

**Reference:**
None
2. Concerning periventricular leukomalacia, which one is CORRECT?

A. Injury is secondary to venous infarction.
B. **Areas of cystic necrosis develop 1-3 weeks after injury.**
C. Infants are at greatest risk for PVL at 36-38 weeks gestation.
D. Ultrasound is sensitive in the identification of the acute lesions.

**Rationale:**
A. Incorrect. Injury is thought to be arterial watershed infarction of the premature infant’s periventricular white matter. Venous infarction is believed to be the mechanism of the parenchymal hemorrhagic component in Grade IV germinal matrix hemorrhage.
B. Correct. The multicystic periventricular components, characteristic at sonography, represent areas of cystic necrosis in ischemic areas, and develop 1-3 weeks after the acute injury, after resolution of the initially echodense lesions.
C. Incorrect. Injury typically occurs in preterm infants less than 32 weeks gestation.
D. Incorrect. Ultrasound is often insensitive in the acute stage, with as many as 50% of initial studies in the first few days following injury reported as normal in appearance.

**Reference:**
Concerning the presence of a perirenal fluid collection in a newborn infant, which one is CORRECT?

A. **This finding is associated with posterior urethral valves.**
B. The finding represents sympathetic transudation of fluid due to obstruction.
C. Secondary infection is not a significant concern.
D. Early post-contrast CT or MR images are diagnostic.

**Rationale:**
A. Correct. Long standing high grade obstruction in utero is associated with renal dysplasia and poor renal function. The presence of loculated or free urine extravasation from the kidney of a newborn infant is most commonly associated with posterior urethral valves, and is secondary to forniceal rupture and decompression into the perirenal space, at times extending into the peritoneal cavity, presenting as urinary ascites. This finding has been associated with relative preservation of function in the kidney, probably due to decompression of the obstruction.
B. Incorrect. This fluid collection results from forniceal rupture of a calyx and extravasation of urine forming a subcapsular urinoma. Subcapsular, perinephric and paranephric urinomas can occur in association with urinary obstruction, including posterior urethral valves, ureteropelvic junction obstruction, neurogenic bladder and bladder tumors or after trauma to the urinary tract. The collection may rupture into the peritoneal space producing urinary ascites.
C. Incorrect. Secondary infection of fluid collections in a newborn is always of concern, especially when there is underlying obstruction and/or reflux.
D. Incorrect. If a contrasted examination (CT or MRI) were done, delayed images would be most useful in demonstrating contrast accumulation within the urinoma, confirming the connection to the urinary tract. In the setting of posterior urethral valves, however, upper tract contrast examinations are unnecessary for diagnosis.

**Reference:**
4. Concerning the thymus in childhood, which one is CORRECT?

A. Hodgkin's disease rarely involves the thymus.
B. Normal thymus obscures the great vessels at sonography.
C. **Thymic cysts may occur in the neck.**
D. Normal thymus is bright on T1- and dark on T2-weighted MR images.

**Rationale:**
A. Incorrect. Hodgkin's disease often occurs in the anterior mediastinum and typically infiltrates the thymus. Bulky adjacent mediastinal adenopathy usually occurs. On CT imaging, the thymus is enlarged, nodular and heterogeneous with areas of necrosis frequently present, as opposed to the smooth homogenous normal thymus. Compression of the airway is often a significant concern.
B. Incorrect. The normal thymus is of uniform, mid-level echogenicity, and offers an excellent window into the mediastinum, by displacing surrounding air-filled lung.
C. Correct. Thymic cysts are most commonly found in thymic rests in the neck, rather than in the main gland. These may present acutely due to infection or hemorrhage. They may be associated with bone marrow aplasia and have been reported in patients with HIV and treated lymphoma.
D. Incorrect. On MR images the normal thymus in childhood is relatively dark (slightly hyperintense to muscle) on T1 weighted images and has uniform bright signal on T2 weighted sequences. In younger infants the thymus has a quadrilateral shape with convex borders, becoming more triangular in children over five years of age. Fatty infiltration of the thymus is usually not seen until the late teenage years or adulthood.

**Reference:**
5. Concerning slipped capital femoral epiphysis, which one is CORRECT?

A. The frog lateral radiograph is contraindicated
B. **Produces knee pain in one quarter of patients**
C. Is bilateral in 75 - 80% of cases
D. Is most common in athletes

**Rationale:**
A. Incorrect. The femoral head slips posteriorly and medially. The frog lateral view is essential to detect the posterior component of the slip, which may be the major component in some patients.
B. Correct. Patients with slipped capital femoral epiphysis usually present with hip or groin pain; however, in approximately 25% of patients pain is referred to the ipsilateral knee. These patients may present with knee pain alone or knee pain in combination with hip/groin pain. In a patient in the typical age range for slipped capital femoral epiphysis, unexplained knee pain should prompt consideration for slipped capital femoral epiphysis.
C. Incorrect. Slipped capital femoral epiphysis is bilateral in approximately one quarter to one third of patients, much less than 75-80%.
D. Incorrect. Slipped capital femoral epiphysis is most common in obese adolescents.

**Reference:**
6. Concerning Pulmonary Sling, which of the following is CORRECT?

A. Left pulmonary artery crosses posterior to the esophagus
B. **Complete tracheal cartilaginous rings increase morbidity**
C. Diverticulum of Kommerell compresses the airway
D. Left ligamentum arteriosum completes the vascular ring

**Rationale:**
A. Incorrect
B. Correct. A pulmonary sling occurs when the left pulmonary artery originates from the right pulmonary artery, crossing the mediastinum from right to left between the trachea and esophagus. A subset of patients may have associated complete tracheal rings leading to significant airway stenosis, increasing morbidity. Diverticulum of Kommerell is associated with aberrant subclavian artery and vascular ring. Pulmonary sling is not a vascular ring and is not dependent on the presence or position of the ligamentum arteriosum.
C. Incorrect
D. Incorrect

**Reference:**
7. Regarding the pediatric duodenum, which one of the following is CORRECT?

A. Duodenal atresia results from in utero vascular accident
B. Duodenal stenosis or atresia is not associated with annular pancreas.
C. Malrotation with volvulus can present with a normal abdominal radiograph.
D. Presurgical diagnosis of duodenal atresia requires UGI.

Rationale:
A. Incorrect. Duodenal atresia, unlike small bowel and colonic atresia, is believed to result from failure of recanalization.
B. Incorrect. In approximately 40% of patients with annular pancreas, there is underlying stenosis or atresia. These patients tend to present earlier, usually in the neonatal period.
C. Correct. Although patients with long-standing or more severe obstruction will tend to have a distended stomach and some distension of the duodenum, with paucity of distal bowel gas, a normal radiograph does not exclude malrotation with volvulus.
D. Incorrect. The diagnosis of duodenal atresia can be made on the plain film findings of double bubble, with distension of the stomach, and marked distension of the duodenum, typically 1/2 to 1/3 the size of the stomach.

Reference:
Concerning enteral duplications, which of the following is CORRECT?

A. Most common at the stomach
B. Most communicate with the adjacent bowel
C. **May act as the lead point in an intussusception**
D. Are best delineated by a barium small bowel follow-through

**Rationale:**
A. Incorrect. The most common location of enteral duplications is in the distal ileum, followed by the esophagus.
B. Incorrect. The majority of duplications do not communicate with the adjacent bowel.
C. Correct. Duplication cysts may act as the lead point of an intussusception, particularly when located in the distal ileum. Duplication cysts may also present with inflammatory changes in cases in which gastric mucosa is present within the cyst, or with segmental volvulus from the mass effect and traction of the cyst.
D. Incorrect. Although contrast studies may show mass effect on bowel by a duplication cyst, they do not typically depict the mass itself. Cross-sectional imaging will better demonstrate the mass and its cystic nature. Ultrasound is the most definitive study, as it will show the cystic nature of the mass and often demonstrate the layered appearance of the cyst wall (inner echogenic mucosa and outer hypoechoic muscle) which is characteristic although not completely specific, of a duplication cyst.

**Reference:**
Concerning Legg-Calve-Perthes disease, which of the following is correct?

A. It occurs more commonly in girls than in boys.
B. Patients usually have bilateral disease at presentation.
C. Contrast-enhanced MRI is not useful in establishing a diagnosis.
D. **Acutely, femoral head marrow is low signal on T1-weighted MRI.**

**Rationale:**
A. Incorrect. Legg-Calve-Perthes disease is more common in boys than girls. The boy to girl ratio is approximately 4:1.
B. Incorrect. Legg-Calve-Perthes disease is usually unilateral. Approximately 85% of cases are unilateral. Bilateral disease is less common (10-15%) and is usually asynchronous at presentation.
C. Incorrect. Administration of intravenous Gadolinium can help to identify viable tissue in areas of bone marrow edema.
D. Correct. MR may show abnormality prior to radiographs and may diagnose Legg-Calve-Perthes disease before radiographic abnormality develops. On MR imaging, the affected femoral head will show decreased signal on T1-weighted images. Normally, the femoral head shows high signal due to fatty marrow. With Legg-Calve-Perthes disease, the femoral head bone marrow becomes edematous, decreasing its signal intensity on T1-weighted images.

**Reference:**
10. Regarding skeletal dysplasias with limb shortening, which of the following is CORRECT?

A. **Rhizomelic - short humerus**
B. Mesomelic - short femur
C. Acromelic - short radius
D. Rhizomelic - short tibia

**Rationale:**
A. Correct. "Rhizomelic" means shortening of the root or proximal bone. The humerus is the proximal bone within the upper extremity.
B. Incorrect. "Mesomelic" means shortening of the middle bone. The femur is not the middle bone of the lower extremity. In the lower extremity, mesomelic refers to shortening of the tibia and fibula.
C. Incorrect. "Acromelic" means shortening of the distal bones. The radius is not a distal bone of the upper extremity. In the upper extremity, acromelic refers to shortening of the hand.
D. Incorrect. "Rhizomelic" means shortening of the root or proximal bone. The tibia is the middle bone of the lower extremity, not the proximal bone. In the lower extremity, rhizomelic refers to shortening of the femur.

**Reference:**