What is the most likely diagnosis?

- Options A and B are NOT correct. This mass is skin covered. Neither a meningomyelocele nor myeloschisis is skin covered.
- Option C is CORRECT. Meningoceles are skin covered neural tube defects.
- Option D is NOT correct. Caudal regression is not associated with a soft tissue mass.
- Option E is NOT correct. The lemon sign is a sonographic sign of hindbrain herniation.

References:
The fetal MRI and US images:  
A. Depict a low grade level in the UTD classification  
B. Represent a high grade UTD  
C. The fetal MRI and US do not correlate with each other  
D. Findings are likely physiologic

The fetal MRI and US images show which of the following findings?  

- Option A is **NOT** correct since a low risk UTD would only include central urinary tract dilatation and not peripheral as in this case.
- Option B is **CORRECT**. The images show high grade UTD. Severe bilateral hydronephrosis with cortical thinning is noted in the coronal fetal MRI image. Similar findings of severe hydronephrosis are seen on the ultrasound images with not significant cortical thinning but minimally increased echogenicity. Despite the gestational age of this fetus, the degree of hydronephrosis and the abnormal cortex increases the risk for renal pathology from low risk to increased risk, UTD A2-3.
- Option C is **NOT** correct because the findings on MRI and ultrasound correlate with each other.
- Option D is **NOT** correct. The findings are not physiologic rather reflect severe bilateral hydronephrosis.

Regarding the abnormalities denoted on this coronal T2-weighted MR image of a 26 week old fetus (See Figure 2 - Arrows), which one of the following is the **TRULY** concerning post-natal clinical outcomes?  

- A. Degree of impairment depends on the location of the cleft, whether it is unilateral or bilateral, and whether there are associated malformations.
- B. Bilateral defects result in less severe neurologic impairment
- C. Ultrasound is better than MRI in accurate prenatal detection and thus can be used to reliably counsel parents about the clinical manifestations
- D. A variety of post-natal therapeutic options exist for dealing with these clefts

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Which of the following best describes this patient’s postnatal clinical outcomes?

The fetal MR image demonstrates bilateral open-lip schizencephalic defects, left larger than right, that are lined by gray matter.

- Option A is **CORRECT**. The severity of motor and mental disability as well as severity of seizure activity is directly related to the extent of the anatomic abnormality and extent of the clefts, respectively.
- Option B is **NOT** correct. Bilateral defects result in worse intellectual and speech development compared with unilateral defects.
- Option C is **NOT** correct. MR imaging is more sensitive than ultrasound in identifying associated brain anomalies and distinguishing schizencephaly from other CSF-containing abnormalities of the fetal brain.
- Option D is **NOT** correct. Currently, no post-natal therapies exist for schizencephaly. Patient care is primarily targeted at seizure control.

Which of the following best describes this patient’s postnatal clinical outcomes?


The images demonstrate measurement of which of the following fetal predictors of CDH severity?

- Option A is **NOT** correct. The lung-head ratio is measured on ultrasound by obtaining the area of the contralateral lung at the level of the 4-chamber view of the heart and dividing by the head circumference
- Option B is **CORRECT**. The method demonstrated in the image is one of the methods used for calculating the total fetal lung volume (TFLV)
- Option C is **NOT** correct. Pulmonary vascularization index is measured on ultrasound using 3-dimensional power Doppler acquisitions
- Option D is **NOT** correct. The modified McGoon index utilizes SSFSE T2-weighted MR images to measure the right and left pulmonary artery and aortic diameters to create a ratio of pulmonary-to-systemic blood flow
- Option E is **NOT** correct. Percent liver herniation also uses consecutive SSFSE T2-weighted MR images and the freehand ROI tool, but the volume of herniated liver is measured and compared to the total liver volume to determine the percentage of liver herniated into the chest
The images demonstrate measurement of which of the following fetal predictors of CDH severity?


Select the CORRECT answer. The findings on this axial image are:

- Option A is NOT correct. The right globe is normal in configuration and size. The left globe is abnormally shaped and small with no lens.
- Option B is NOT correct. Persistent hyperplastic primary vitreous can be unilateral or bilateral. On fetal MRI image, the affected globe(s) is/are small, the lens(es) is/are thick and irregular and there is a dark T2 band attached to the posterior surface of the lens(es).
- Option C is NOT correct. The eyes are widely spaced, not too close together.
- Option D is CORRECT. The left globe is small (microphthalmia) and the fetal eyes are widely spaced (hypotelorism). Microphthalmia can affect one or both globes.
- Option E is NOT correct. The posterior fossa and 4th ventricle are included on this image and are normal in appearance.

Select the CORRECT answer. The findings on this axial image are:

Which one of the following is the MOST likely diagnosis based on this MRI performed at 27 weeks?

Fetal MRI shows a large prepontine and premesencephalic cyst causing compression and displacement of the brainstem, consistent with an arachnoid cyst.

- Option A is NOT correct. A Blake pouch cyst would be centered in the cistern magna or the cerebellomedullary angle.
- Option B is NOT correct. There is compression and displacement of the brainstem related to an extra-axial cyst. Although underdevelopment of the brainstem may be present, mass effect accounts for the dysmorphology.
- Option C is NOT correct. The cerebellum is normal.
- Option D is CORRECT. A large midline cyst compresses the brainstem and displaces it posteriorly. This is consistent with an arachnoid cyst, probably affiliated with the membrane of Liliequist.
- Option E is NOT correct. Joubert syndrome manifests with a malpositioned, hypoplastic vermis, apposed cerebellar hemispheres, and dysmorphic midbrain demonstrating a "molar tooth malformation".

Which one of the following is the MOST likely diagnosis based on this MRI performed at 27 weeks?

Select the correct answer in reference to facial malformations.

- Option A is NOT correct. Hypotelorism may be seen in the setting of craniosynostosis, chromosomal defects such as trisomy 13 and with abnormal brain development, frequently in the spectrum of holoprosencephaly.
- Option B is NOT correct. Hypotelorism is frequently seen in the spectrum of holoprosencephaly.
- Option C is CORRECT. Hypotelorism is frequently seen in the spectrum of holoprosencephaly.
- Option D is NOT correct.

The SSFSE T2-weighted MR image shows which ONE of the following imaging features of a hernia sac in congenital diaphragmatic hernia?

- Option A is CORRECT. The coronal image shows lung superior to the hernia.
- Option B is NOT correct. The image is in the coronal plane. One would expect to see lung posterior to the hernia contents on an axial image, but no axial image is provided.
- Option C is NOT correct. Although having ascites within the hernia sac and separated from the pleural space is associated with the presence of a hernia sac, that finding is not shown in the image.
- Option D is NOT correct. Pleural fluid kept from mingling with the hernia contents would be a sign of a hernia sac if it was present, but pleural fluid is not shown on the provided image.
- Option E is NOT correct. Having free fluid contiguous between the pleural space and the hernia contents would be evidence against a hernia sac.
The SSFSE T2-weighted MR image shows which ONE of the following imaging features of a hernia sac in congenital diaphragmatic hernia?


Which diagnosis is most likely to be seen prenatally?

- Options A and B are NOT correct. Hirschsprung disease and meconium ileus are often a surprise diagnosis.
- Option C is CORRECT. ARM/cloaca have a connection to the GU tract and therefore there is more fluid in the intestines, an abnormally short meconium column on MRI, and mixing of urine and meconium creating calcifications.
- Option D is NOT correct. Ileal atresia often occurs late in gestation.
- Option E is NOT correct.

Which diagnosis is most likely to be seen prenatally?

Axial T2-weighted image from a fetal brain MRI at 28 weeks gestation (See Figure 1) and post-natal follow-up image from the same child (See Figure 2) are provided. What is the diagnosis in this patient?

A. Normal for gestational age
B. Polymicrogyria
C. Lissencephaly
D. Schizencephaly
E. Holoprosencephaly

What is the diagnosis in this child imaged at 28 weeks gestation (A) and postnatally (B)?

• Option A is NOT correct. The normal brain is still smooth in the early 2nd trimester though, by 28 weeks gestation, normal convexity sulci should be seen.
• Option B is NOT correct. Polymicrogyria is characterized by excessively small gyri and prominent convolutions with altered sulcation pattern.
• Option C is CORRECT. Lissencephaly is a malformation of cortical development caused by arrested neuronal migration resulting in a smooth brain. On imaging, 3 layers are seen: a thin outer cortical layer of neurons, a “cell-sparse” zone, and a thick deep cortical layer of neurons.
• Option D is NOT correct. Schizencephaly is a trans-mantle cleft of the brain lined by gray matter.
• Option E is NOT correct. Holoprosencephaly is a complex malformation characterized by abnormal cleavage of the forebrain (prosencephalon).

You are shown an image from a prenatal MR (coronal T2 SSFSE) of a 22 week old fetus. What is the MOST LIKELY diagnosis?

A. CPAM
B. Bronchogenic Cyst
C. Congenital lobar overinflation
D. Bronchopulmonary sequestration
E. Hybrid Lesion

What is the diagnosis in this child imaged at 28 weeks gestation (A) and postnatally (B)?

What is the MOST LIKELY diagnosis on this prenatal MR of a 22 week old fetus?

- Option A is NOT correct. CPAM is not the best answer. Although cysts are present in the mass, a feeding vessel is not characteristic of a CPAM.
- Option B is NOT correct. Bronchogenic cyst is not correct. These typically appear as T2 hyperintense structures on MR and are classically located in a subcarinal region. No feeding vessel or solid tissue is evident in these.
- Option C is NOT correct.
- Option D is NOT correct. Although a bronchopulmonary sequestration would demonstrate a feeding vessel, cysts would not be expected in these masses.
- Option E is CORRECT. A hybrid lesion is the best answer. This lesion has components of both a CPAM and a bronchopulmonary sequestration compatible with a hybrid lesion.

Sagittal (Figure 1a) and axial (Figure 1b) T2-weighted images from a fetal MR at 22 gestational weeks are shown. Which one of the following is the MOST likely diagnosis?

A. Classical Dandy-Walker Malformation
B. Dandy-Walker continuum (Hypoplasia of the vermis with rotation)
C. Arachnoid Cyst
D. Mega cisterna magna
E. Joubert syndrome

Which of the following entities is the MOST likely diagnosis in this fetus of 22 weeks gestational age?

- Fetal MRI shows 4th ventriculomegaly, marked counter-clockwise rotation of a hypoplastic vermis with an increased tegmento-vermian angle, and splaying of the cerebellar hemispheres. The torcular herophili is normal in position.
- Option A is NOT correct. Classical Dandy-Walker Malformation is defined by the following criteria: counter-clockwise rotation of a hypoplastic vermis; marked 4th ventriculomegaly; and enlargement of the posterior fossa and torcular elevation.
- Option B is CORRECT. Imaging findings meet all criteria for classical Dandy-Walker Malformation except elevation of the torcular herophili. Therefore, this represents a Dandy-Walker continuum.
- Option C is NOT correct. An arachnoid cyst can cause mass effect and distortion of the regional brain parenchyma, but it is not typically associated with cerebellar hypoplasia.
- Option D is NOT correct. A mega cisterna magna does not splay the cerebellar hemispheres, malnurrate the vermis, or cause vermian hypoplasia.
- Option E is NOT correct. Although Joubert syndrome may manifest with a malpositioned, hypoplastic vermis, the cerebellar hemispheres are typically apposed and the midbrain is dysmorphic, with thinning of the lower tegmentum and thickened superior cerebellar peduncles ("molar tooth malformation").
Which of the following entities is the MOST likely diagnosis in this fetus of 22 weeks gestational age?


Which of the following is an exclusion criteria for fetal meningomyelocele surgery?

- A. Meningomyelocele at L3
- B. Normal karyotype
- C. Presence of hindbrain herniation
- D. Presence of club feet
- E. Scoliotic curvature of > 30°

Which of the following is an exclusion criteria for fetal meningomyelocele surgery?

- Options A, B, and C are NOT correct. They are inclusion criteria for surgery
- Option D is NOT correct. Club feet do not exclude a patient from undergoing fetal surgery
- Option E is correct. Severe scoliosis has been used as an exclusion criteria for fetal surgery

What was one of the purposes of the new UTD classification?

A. Create more confusion amongst the pediatricians
B. Create more confusion amongst the pediatric radiologists
C. Unify the language used by MFMs and pediatric radiologists only
D. Standardize terminology of UTD that could be applied both prenatally and postnatally
E. All of the above

What was one of the purposes of the new urinary tract dilatation classification?

Which of the following is true regarding the abnormality shown on this ultrasound of the chest in a fetus imaged at 32 weeks of gestation?

- Option A is NOT correct. CPAMs grow most rapidly during the 20-26 weeks of gestation after which growth peaks and plateaus.
- Option B is NOT correct. CPAMs typically have abnormal communication with the tracheobronchial tree.
- Option C is CORRECT. Fetal hydrops is an indicator of poor prognosis in patients with congenital pulmonary airway malformation.
- Option D is NOT correct. CPAMs have pulmonary arterial supply.
- Option E is NOT correct. Immediate postsurgical resection is not indicated in asymptomatic, infants. Surgical resection may be performed in symptomatic neonates.

Which is true about distal intestinal obstruction?

- Option A is NOT correct. Echogenic bowel is very common in 1.8% of all second trimester fetuses.
- Option B is NOT correct. Because the fluid is absorbed in patients with a distal intestinal obstruction, polyhydramnios is not expected.
- Option C is CORRECT. Distal bowel obstructions are not well detected prenatally – this is because they either occur late in gestation and/or there is a normal amount of amniotic fluid.
- Option D is NOT correct. Calcifications are only be seen in the intestines in patients with an anorectal malformation or cloaca.
- Option E is NOT correct.
Which is true about distal intestinal obstruction?