Advances in IUGR/Fetal Distress Assessment

Wesley Lee, MD

1. Which of the following prenatal imaging parameters best predicts neonatal nutritional status based on percentage body fat?
   A. estimated fetal weight
   B. fetal abdominal circumference
   C. fetal fractional thigh volume
   D. fetal umbilical artery Doppler ultrasound

Correct Answer: C

Rationale
We have traditionally measured fetal skeletal parameters using conventional 2D US (e.g. biparietal diameter, head circumference, femur diaphysis length) to predict neonatal growth outcome. Abdominal circumference does include some soft tissue but, mainly reflects liver size. Fractional thigh volume is defined by both limb soft tissue and long bone length. Pediatric air displacement plethysmography indicates that fractional thigh volume is a much better fetal parameter than obtained by traditional 2D biometry for the prediction of percentage body fat in newborns.

References

2. Estimated fetal weight by conventional 2D methods is associated with which of the following typical error ranges?
   A. within 10 percent
   B. within 15 percent
   C. within 20 percent
   D. within 25 percent

Correct Answer: B

Rationale
Melamed and colleagues (2009) compared the accuracy of 26 different birth weight prediction models using 3,705 sonographic weight estimations less than 3 days before delivery. For most models, the estimates were within 15% of the actual BW in greater than 80% of the cases.

Reference
3. According to the PORTO study, which of the following pair of prenatal parameters were strongly and most consistently associated with adverse perinatal outcome?
   A. abnormal uterine artery Doppler and EFW < 5th percentile
   B. abnormal umbilical artery Doppler and EFW < 3rd percentile
   C. abnormal middle cerebral artery Doppler and EFW < 10th percentile
   D. abnormal ductus venosus Doppler and EFW < 10th percentile

Correct Answer: B

Rationale
The PORTO study was a prospective observational multi-center study of over 1100 consecutive ultrasound dated pregnancies that were complicated by abnormally low estimated fetal weight. Abnormal umbilical artery Doppler and EFW < 3rd centile were strongly and most consistently associated with adverse perinatal outcome.

Reference

Fetal Supratentorial Abnormalities: Case Based
Orit A. Glenn, MD

4. Which of the following statements about polymicrogyria on fetal MRI is true?
   A. It appears as too few infoldings of cortex.
   B. It is rarely occurs with schizencephalies.
   C. It can occur following in utero injury.
   D. It is best diagnosed prior to 20 weeks gestation.
   E. It is always genetic in etiology.

Correct Answer: C

Rationale
Andert C is correct. Polymicrogyria can occur following injury to the developing cortex, such as following an infarct. Answer A is wrong: Polymicrogyria appears as too numerous infoldings of the cortex, and do not correspond to normal sulci. Answer B is wrong. Polymicrogyria is frequently seen lining the clefts of a schizencephalic defect. Answer D is wrong: Polymicrogyria, and other malformations of cortical development, are difficult to diagnose in a young fetus, related to fetal motion and small size of structure being imaged, and are more easily diagnosed at later gestational ages.

References
5. Which of the following is FALSE about the corpus callosum on fetal MRI?
   A. In cases of suspected callosal hypogenesis, the callosal length should be measured on midline sagittal image.
   B. Anterior columns of fornices can be mistaken for the cavum septum pellucidum.
   C. Thin corpus callosum is difficult to diagnose by fetal MRI.
   D. Callosal agenesis is usually isolated on fetal MRI.
   E. Absent septal leaves can be an indication of callosal agenesis.

Correct Answer: D

Rationale
Answer D is correct. Callosal agenesis is frequently seen with other malformations, most commonly cortical malformations. Therefore, it is importantly to carefully look for other abnormalities when callosal agenesis is diagnosed by fetal MR. Answer A is wrong: The callosum should always be measured on a midline sagittal image, and compared with normative data when callosal hypogenesis is considered. Answer B is wrong: Because the anterior columns of the fornices can contact each other, they can mimic the cavum septum pellucidum, although they can be distinguished from the cavum due to presence of a midline echogenic structure. Answer C is wrong: The body of the callosum normally measures 1-2mm in thickness, making it difficult to detect a thin corpus callosum. Answer E is wrong: The septal leaves fail to form when there is callosal agenesis.

References

Fetal Posterior Fossa Abnormalities: Case Based
Ashley Robinson, MB ChB, FRCR, FRCPC

6. Key morphologic landmarks that indicate the vermis is present on midline sagittal images include all of the following except:
   A. The primary fissure
   B. The fastigium (fastigial point)
   C. The obex
   D. The secondary fissure
   E. The declive

Correct Answer: C

Rationale
The obex is the inferior recess of the fourth ventricle and not part of the vermis.


References


7. At 22 weeks menstrual age the following morphologic findings indicate a good prognosis except:
   A. A vermis with multiple lobules visible
   B. A vermis with an anterior to posterior lobe ratio of approximately 50%
   C. A vermis that is angulated away from the tegmentum but with no other morphologic abnormality
   D. A vermis where no primary fissure is visible
   E. A vermis with a cranio-caudal diameter of 11mm

Correct Answer: D

Rationale
The primary fissure should always be visible after about 17.5 weeks gestation.

References


ii. Robinson AJ, Goldstein RG. The cisterna magna septa - a vestigial remnant of Blake’s pouch and a potential new marker for normal development of the rhombencephalon. J Ultrasound Med. 2007; 26: 83-95


8. Choline is a metabolite that can be identified using proton MR spectroscopy in the fetal brain: Which of the following statements is incorrect:
   A. Choline resonates at 3.2 ppm
   B. Choline is a neuronal marker
   C. Choline plays a role in myelination
   D. Choline declines with increasing gestational age

Correct Answer: B
Rationale
N-acetylaspartate is a neuronal marker

References
i. Limperopoulos, C. Magnetic Resonance Spectroscopy of the fetal brain. in MR Spectroscopy of Pediatric Disorders, Bluml and Panigrahy, eds. Springer 2013

9. Which of the following brain tissues demonstrates the greatest maturational rate in the third trimester of pregnancy by quantitative MRI?
   A. Cortical gray matter
   B. White matter
   C. Brainstem
   D. Ventrices

Correct Answer: A

Rationale
A more rapid increase in cerebral gray matter volume occurs compared to white matter in the third trimester. Major phases of cerebral hemispheric white matter development (axonal outgrowth and myelination) occur later in the third trimester and beyond.

References

What Does the Neurologist Want to Know From the Fetal MRI?
Terrie E. Inder, MD, PhD

10. Fetal MRI:
   A. Is a complementary neuroimaging technique to ultrasound in high risk settings
   B. Does not require specialized sequences on MRI scanner
   C. Can be easily interpreted without knowledge of infant’s gestation
   D. Has low but proven risk to the fetus from scanning

Correct Answer: A

Reference
11. Fetal MRI:
   A. Is best performed on a high field (≥ 3T) MRI scanner
   B. Can detect additional pathology and diagnoses in > 75% of cases
   C. Is best performed at 14-18 weeks gestation
   D. Can yield a superior diagnostic yield than ultrasound in cortical malformations

Correct Answer: D

Reference

Relevance of Placental Findings to Fetal Brain Injury and Ultimate Neuro Outcome
Jeffrey J. Neil, MD, PhD

12. Of the following, which does not contribute to the challenges of associating placental pathology with neurodevelopmental outcome?
   A. Most placentas are discarded
   B. Developmental impairment may not be manifest for years
   C. Measures of placental pathology are not standardized
   D. Samples of convenience are often used
   E. Developmental impairments are common

Correct Answer: E

Reference

Neonatal Cranial US Update: Case Based
Fred E. Avni, MD, PhD

13. What would be a typical site for brain hemorrhage or ischemic damage demonstrated by US in at-term neonate?
   A. Periventricular areas
   B. Cortico-subcortical brain layer
   C. Lateral ventricles
   D. Third ventricle
   E. Corpus callosum

Correct Answer: B

Rationale
A - The periventricular areas are less vulnerable than in premature thanks to progressive brain maturation.
B - This is one of the most typical area of hypoxic or hemorrhagic insult in neonates at terms in relation with venous infarcts. The other potential sites are the peri-sylvian areas or the entire supra-tentorial brain. C- Lateral ventricles – incorrect. D - Third ventricle – incorrect. E - Most unusual area of damage
14. What US finding carries a poor prognosis in terms of permanent sequelae?
   A. Faint hemorrhage in the lateral ventricles
   B. Peri-cerebral effusion
   C. Subependymal hemorrhage
   D. Bright brain pattern
   E. Transient periventricular hyperechogenicity

Correct Answer: D

Rationale
A - Usually a transitory finding with no developmental consequences
B - Can be found as incidental finding. No consequences if isolated and without parenchymal damage
C - Unusual in term neonates; if isolated, the prognosis is good
D - Bright Brain pattern results from a diffuse hypoxic insult to a large part of the supra-tentorial brain. This carries a poor developmental prognosis with motor sequelae.
E - This is unusual in term neonates. If it occurs, it is usually transient without consequences

References
i. Dinan D Daneman A Guimaraes CV & al Easily overlooked US findings in the evaluation of neonatal encephalopathy: lessons learned from MRI  Semin Ultrasound CT MR  2014; 35 (6) 627-651

15. Which of the following is NOT true about white matter injury of prematurity?
   A. Timing of white matter injury is related to developmental peak of subplate zone
   B. It is the most common type of CNS injury in the preterm neonate.
   C. It is due to susceptibility of pre-myelinating oligodendrocytes and subplate neurons to injury
   D. It can be focal or diffuse
   E. Diffuse white matter injury is better detected with US than MRI.

Correct Answer: E
Rationale
Answer E is correct: Whereas cystic white matter injury can be diagnosed by ultrasound, diffuse white matter is not well diagnosed by ultrasound. Answer B and D are wrong: White matter injury of prematurity is a common type of brain injury in preterm neonates, and can be both focal and diffuse, and cystic and noncystic. Answer A and C are wrong: The selective vulnerability of pre-OL cells and subplate neurons have an important role in white matter injury of prematurity, as the peak timing of white matter injury of prematurity overlaps with the developmental period of the subplate zone.

References

16. Regarding periventricular hemorrhagic injury, which is the most accurate?
   A. It results from a direct extension of hemorrhage from the germinal matrix into the parenchyma.
   B. It rarely has a poor prognosis.
   C. It may be associated with nonhemorrhagic ischemia
   D. It is an arterial infarct.
   E. It is only seen in preterm neonates

Correct Answer: C

Rationale
Answer C is correct: It is a venous infarct resulting from compression of medullary veins, and thus can have both hemorrhagic and nonhemorrhagic ischemic changes. Answers A and D are wrong: It is a venous, not arterial, infarct. It is not the result of direct extension of germinal matrix hemorrhage. Answer B is wrong: It is associated with a poor prognosis, which depends on location and extent of PVHI. Answer E is wrong: it can occur in utero, and therefore can be seen in children born at term.

References

The Predictive Value of MRI in the Premature Infant
Terrie E. Inder, MD, PhD

17. Magnetic Resonance Imaging at term equivalent in the preterm infant has:
   A. Improved sensitivity for cystic periventricular leukomalacia
   B. High positive predictive value of no abnormality for normal outcome
   C. Detection of abnormalities of significance in >50% of premature infants
   D. Requires sedation to enable high quality imaging to be acquired
   E. Improved prediction by inclusion of Diffuse Excessive High Signal Intensity characteristics

Correct Answer: B
References


iii. Smyser CD, Kidokoro H, Inder TE. Magnetic resonance imaging of the brain at term equivalent age in extremely premature neonates: to scan or not to scan? J Paediatr Child Health. 2012 Sep;48(9):794-800


18. Which of the following abnormalities on an MRI at term is not of any prognostic significance?
   A. Diffuse High Excessive High Signal Intensity on T2 weighted imaging
   B. Myelination in the Posterior Limb of the Internal Capsule
   C. Low brain volume
   D. Enlarged interhemispheric distance

Correct Answer: A

References


iii. Smyser CD, Kidokoro H, Inder TE. Magnetic resonance imaging of the brain at term equivalent age in extremely premature neonates: to scan or not to scan? J Paediatr Child Health. 2012 Sep;48(9):794-800

Fetal Spine: Case Based
Dorothy I. Bulas, MD, FACR

19. Which of the following are inclusion criteria for possible fetal surgery?
   A. Meningomyelocele above S1
   B. Normal Karyotype
   C. Presence of hindbrain herniation
   D. Singleton pregnancy from 19 to 25 weeks 6 days
   E. All of the above

Correct Answer: E

References


Rationale
In utero repair of open neural tube defects (ONTD) is now being performed in selected Patients. Fetuses with ONTD develop neurologic deficits by the primary neurulation defect that creates the dysraphism and associated myelodysplasia. It is thought that further injury occurs from chemical and/or physical trauma to the neural tissue as a result of exposure to amniotic fluid. Fetal surgery could reduce this later injury by covering the exposed spinal cord. Studies on animals and human fetuses suggested some improvement in distal sensorimotor function. In addition, reversal of hindbrain herniation with decreased need for ventricular shunting has been noted. The multicenter randomized prospective National Institute of Health trial (MOMS) convincingly showed that open fetal surgery could improve neurologic outcome with decreased need for shunting. Accurate diagnosis is critical. All the criteria listed are inclusion criteria required for fetal surgery.

20. Which of the following is true concerning this 20 week gestation with the lemon sign?
   A. Meningomyelocele is present
   B. AFP is elevated
   C. Chiari malformation is present
   D. All of the above
   E. Cannot tell

Correct Answer: E

References

Rationale
Concavity of the frontal bones, the “lemon sign” can be seen in 98% of cases of ONTD before 24 weeks. It has been described as early as 13 weeks’ gestation. The “lemon sign”, however, is not a sign specific to ONTD, and is present in up to 1% to 2% of normal fetuses. It can be associated with occipital meningoceles and encephaloceles as well. The skull configuration typically resolves by the third trimester in fetuses with ONTD (present in only 13% of affected fetuses after 24 weeks).

Updates on CNS Fetal Surgery: MOMS
Darrell L. Cass, MD

21. Which of the following is the least likely therapeutic benefit to antenatal repair of an open neural tube defect?
   A. Decreased need for a ventriculoperitoneal shunt.
   B. Increased gestational age at birth.
   C. Decreased hindbrain herniation.
   D. Improved ability to walk independently at 30 months of life
   E. Decreased brainstem kinking.

Correct Answer: B
Rationale:
Answer is b. The MOMS trial published in the NEJM in March, 2011 compared outcomes of patients treated with open fetal surgery repair of myelomeningocele to those treated with postnatal repair. Outcomes were reported from 78 fetuses randomized to fetal repair (77 had the fetal repair) in comparison to 80 who had postnatal repair and included the gestational age at birth, fetal and postnatal mortality, need for a ventriculoperitoneal shunt by 1 year of age, and motor and cognitive testing at 30 months of age. Those who had fetal repair were born earlier than those who had postnatal repair (34.1 vs 37.3 weeks). However, those who had fetal repair demonstrated half the need for a VP shunt (40% vs 82%) at 1 year of age and twice the ability to walk independently at 30 months (42% vs 21%). One of the therapeutic benefits to fetal repair is reversal of hindbrain herniation. In the results of the MOMS study, 96% of those who had postnatal repair showed any degree of hindbrain herniation at 1 year of age, compared to 64% in those who had fetal repair. A decrease in any degree of brainstem kinking was also noted (20% in those who had fetal repair, compared to 48% of those repaired postnataally).

22. Which of the following is the highest risk associated with open fetal surgery repair of an open neural tube defect?
   A. Fetal demise
   B. Placenta accreta in future pregnancies
   C. Need for maternal blood transfusion
   D. Preterm birth
   E. Periventricular leukomalacia

Correct Answer: D

Rationale
The MOMS study demonstrated that the highest risk associated with open fetal surgical repair was preterm birth. In that study, 85% of fetuses who had postnatal repair were born at 37 weeks’ or later, whereas only 21% of fetuses who had fetal repair were born at 37 weeks or later. Any hysterotomy may pose a risk for placenta accreta in future pregnancies; however the incidence of placenta accreta following open fetal surgery is unknown. Likely the risk is low as no cases have been reported to date. In the MOMS study, those mothers who underwent fetal repair had an increased risk of blood transfusion at delivery (9% vs 1%). In the MOMS study, 2 of 77 (3%) fetuses who underwent fetal repair died in utero, whereas no fetus who did not have fetal repair died antenatally.

Reference

Neonatal Spine US: Case Based
Leann E. Linam, MD

23. All of the following are characteristics of a normal spine US except:
   A. Pulsatility of nerve roots
   B. Filum resting halfway between anterior and posterior walls of spinal canal
   C. Conus L3-4
   D. Filum terminale 2mm or less

Correct Answer: C
24. This ultrasound was performed for sacral dimple. Which of the following is true?
   A. There is a cyst of the filum terminale.
   B. The conus is low-lying.
   C. There is a presacral mass.
   D. There is a lipomyelocele.
   E. Spinal dysraphic defect is seen.

Correct Answer: B

References

Neuroimaging after HIE: What are the Latest Recommendations from Our Clinicians?
Jeffrey J. Neil, MD, PhD

25. Which of the following is the most common pattern of injury found on MRI of encephalopathic, term-born infants?
   A. Basal ganglia-thalamic injury
   B. Arterial distribution infarct
   C. Brainstem injury
   D. Periventricular white matter injury
   E. Global injury

Correct Answer: A

References

26. Which of the following is most strongly predictive of motor outcome?
   A. Presence of watershed injury
   B. Involvement of the PLIC
   C. Presence of ventriculomegaly
   D. Involvement of the occipital lobe
   E. Presence of extra-axial blood

Correct Answer: B

References


27. Select the correct statement regarding lymphatic malformations:
   A. May arise in the posterior or anterolateral neck
   B. Non-nuchal lymphatic malformations are highly associated with chromosomal anomalies
   C. Nuchal LM are usually seen in the first trimester
   D. A and C
   E. B and A

Correct Answer: D

Reference

28. Which statement is false?
   A. Epulis arise from the gums of the alveolar ridge
   B. Sacral teratomas are more frequent than oral and neck teratomas
   C. Amniotic fluid AFP is very helpful in the diagnosis of cervical teratomas
   D. Intra-amniotic injection T4 is one of the treatments for fetal goiter
   E. The most common type of branchial cleft cyst is type 2

Correct Answer: C

Reference

The Many Facets of VACTERL: Case Based
Teresa Chapman, MD, MA

29. How many core anomalies are required for the diagnosis of VACTERL?
   A. Three
   B. Four
   C. Five
   D. Six
   E. Seven

Correct Answer: A

Rationale
By definition, the VACTERL association requires three core anomalies (vertebral anomaly, anorectal malformation, cardiac anomaly, tracheoesophageal fistula, esophageal atresia, renal anomaly, limb anomaly) to establish this diagnosis. The VATER association was first described in 1973. Subsequently in 1974, Temtamy and Miller expanded the condition to include cardiac defects, and then in 1975, Kaufman et al. changed the acronym to VACTERL to include cardiac malformations and limb anomalies.
References

30. Which of the following renal anomalies is the most frequently seen in VACTERL/VATER cases?
A. Moderate pyelectasis
B. Cystic dysplasia
C. Unilateral agenesis
D. Duplex collecting system

Correct Answer: C

Rationale
Although vesicoureteral reflux is seen in approximately a quarter of patients with VACTERL, the most commonly observed renal structural anomaly is renal agenesis, seen in approximately 25%. Other renal malformations observed include cystic dysplasia, isolated hydronephrosis, horseshoe kidney, renal hypoplasia, and renal ectopia.

References

31. Which of the following findings is more readily assessed by fetal MRI than by sonography in the diagnosis of TEF/esophageal atresia?
A. Absent stomach
B. Esophageal pouch
C. Abnormal trachea
D. Polyhydramnios

Correct Answer: B

Rationale
Sonographic findings that raise suspicion for this diagnosis include polyhydramnios and small or absent stomach. At best, this combination of findings only offers a positive predictive value of 56%. The differential considerations for either of these findings alone are quite extensive. Fetal MRI allows one to exploit the obvious, hyperintense fluid signal within a distended atretic esophageal pouch. Because of the outstanding tissue differentiation provided by MRI, prenatal recognition of upper esophageal pouches is more readily done by MRI than by sonography.

Reference
**Fetal Lung Masses: Case Based**  
Fred E. Avni, MD, PhD

### 32. About broncho-pulmonary malformation, what statement is correct?

A. Prenatal US represents the best method to evaluate the extent of a CPAM  
B. fMRI is the best method to evaluate the extent of a CPAM  
C. Post natal Chest CT is best method to evaluate the extent of a CPAM  
D. US + Color Doppler is the best method to evaluate the extent of a CPAM.  
E. Post natal Chest Xray is the best method

**Correct Answer: C**

**Rationale**  
A - No, prenatal US cannot differentiate malformed pulmonary tissue from an area with distended alveoli filled with fluid due to compression. Therefore, US overestimates the lesion.  
B - fMRI can better than US evaluate the extent of a CPAM, but also, it might be difficult to differentiate malformation from areas appearing hypersignal due to bronchial compression.  
C - Clearly, postnatal chest CT is the best method to differentiate normal overdistended lung from lung involved by malformation. Associated systemic vessels and venous return are also more easily demonstrated if present.  
D - Same answer than in a). Still color Doppler may be helpful in demonstrating areas with normal or abnormal lung vascularization.  
E - Post natal chest Xray underestimates the remaining malformation but can demonstrate hyperinflation.

**References**  
i. Riedling WFJ Vargas SO Jennings RW & al Bronchial atresia is common to ELS, ILS, CCAM and LE Pediatr Dev Pathol 2006; 9: 361-373  
iii. Epelman M Kreiger PA Servaes S & al Current imaging of prenatally diagnosed congenital lung lesions Semin Ultrasound CT MRI 31: 141-157  

### 33. One of the following diagnosis should not be considered in case of the detection of an echogenic right thoracic mass containing visible cysts:

A. CPAM  
B. hybrid mass (sequestration + CPAM)  
C. Sequestration  
D. Bronchial atresia  
E. Pneumoblastoma

**Correct Answer: D**

**Rationale**  
A - This is a typical aspect of a CPAM, so it should be considered.  
B - Hybrid masses can mix different echographic patterns and appear as echogenic mass with large cysts.  
C - This would be unusual, but sequestration may appear as echogenic mass containing cysts.  
D - In bronchial atresia, the main finding is a mass consisting of fluid filled alveoli; the mass is echogenic and homogenous without cysts.  
E. This rare perinatal pulmonary tumor contains echogenic areas mixed with cystic lesions.
Fetal Neck and Chest Surgical Management Updates
Darrell L. Cass, MD

34. You are shown a fetal MRI of a fetus with a large neck mass. When reviewing the fetal MRI, which of the following is associated with a lower risk for life-threatening airway obstruction at birth?
A. Imaging features which suggest a lymphatic malformation diagnosis.
B. The finding of polyhydramnios
C. Imaging features that suggest a teratoma diagnosis.
D. A tracheoesophageal displacement index (TEDI) of 20

Correct Answer: A

Rationale
Answer is a. Prenatal imaging findings in fetuses with large neck masses that suggest airway obstruction include the finding of polyhydramnios (which suggests esophageal compression and therefore compression of the tracheoesophageal complex) and the diagnosis of a teratoma, as opposed to a lymphatic malformations. Whereas large lymphatic malformations may lead to fetal airway compression and obstruction, the solid nature and rapid growth characteristics of teratomas raise airway obstruction concerns.

The tracheoesophageal displacement index, or TEDI, described by Lazar and colleagues at Texas Children's Hospital in Houston, Texas, is an index which helps measure the degree of displacement of the tracheoesophageal complex away from its usual anatomic location. In the report from Lazar, the TEDI was shown to correlate with the degree of airway displacement and obstruction at birth and the associated benefit of an EXIT delivery.

References
35. Which of the following clinical scenarios is associated with the lowest risk of perinatal survival?

A. A 22-week fetus with a left-sided, non-cystic, lung malformation, with CCAM-volume ratio (CVR) of 2.5.
B. A 26-week fetus with a right-sided, cystic lung malformation, CVR 3.0 and the finding of moderate ascites.
C. A 26-week fetus with a homogeneous, right-sided lung malformation, dilated right mainstem bronchus, and absence of normal right fetal lung, suggesting a diagnosis of mainstem bronchial atresia.
D. A 24-week fetus with cystic right-sided lung malformation, CVR 2.8 and signs of fetal hydrops.
E. A 24-week fetus with solid left-sided lung malformation, CVR 4.5 with signs of fetal hydrops and early cardiac failure who undergoes attempted open fetal surgical resection.

Correct Answer: C

Rationale
Answer is c. Mainstem bronchial atresia is a very rare entity that is associated with 100% mortality currently. This diagnosis should be considered when fetal MRI shows a very large, homogeneous right-sided lung malformation with involvement of all 3 right lung lobes and signs of a dilated right mainstem bronchus. Authors hypothesize the fetal surgical excision of the affected right lung prior to the onset of advance fetal hydrops may provide the only hope for survival, though attempts thus far have been unsuccessful.

Factors associated with worse outcomes for fetuses with lung malformations include a CCAM-volume ratio, or CVR, > 1.6, the findings of fetal hydrops and echocardiographic findings of evolving heart dysfunction, such as absent or reversed flow in the ductus venosus in diastole, worsening tricuspid regurgitation and evolving mitral regurgitation. Of these predictors of outcomes, investigators at Texas Children’s Hospital have shown that the echocardiographic findings of heart failure have the highest predictive value of perinatal demise. Open fetal surgery resection of fetal lung malformations in the setting of fetal hydrops and early cardiac dysfunction is associated with 50-75% survival.

References

Congenital Diaphragmatic Hernia: Cased-based Fetal Imaging
Christopher I. Cassady, MD

36. Which “expected value” reference tables should be used when estimating observed/expected LHR on ultrasound for the fetus in this image?

A. Left CDH (measuring right LHR)
B. Right CDH (measuring left LHR)
C. Neither, because there are bilateral defects
D. Neither, because such tables don’t exist
E. LHR? You are crazy. There is unlikely to be pulmonary hypoplasia.

Correct Answer: E
37. Relatively, the fetus in this image might be considered to have imaging findings favoring a better outcome because there is:
   A. Liver down
   B. a VSD
   C. Lung capping
   D. Spleen capping
   E. no PA hypertension

Correct Answer: C

38. Current consideration for tracheal occlusion therapy (FETO) in utero for severe pulmonary hypoplasia is limited to candidates with:
   A. Mediastinal hernia
   B. Right-sided intrapleural hernia
   C. O/E LHR < 25%
   D. Liver down

Correct Answer: C
39. When differentiating a congenital diaphragmatic hernia (CDH) and congenital pulmonary airway malformation (CPAM) on chest x-ray, which statement is true?

A. Congenital diaphragmatic hernias always demonstrate increased number of air filled cysts in the chest when compared to CPAM.
B. The location of the stomach is always helpful in discriminating CPAM and CDH.
C. The initial first x-ray after birth prior to line placement is usually not helpful when discriminating a CPAM and CDH.
D. The air cysts in CDH are uniform in size whereas the CPAM cysts are variable.

Correct Answer: C

Rationale
A: CDH can have variable amounts of herniated bowel with a potential for more or less air filled bowel loops. CPAMs may have no cysts, 1 cyst or innumerable cysts. B: The gastric location is typically helpful in discriminating CPAM and CDH; however, not always as 10% of left CDH and almost 100% of right sided CDH have an intraabdominal stomach. C: Both CPAM and CDH on initial x-ray will appear radiopaque before air is introduced into the viscera or CPAM cysts from the tracheobronchial tree. D: Although CDH air filled cysts are often uniform, the gastric bubble is variable in distention and sometimes dilated loops will develop. In general, though CPAM cysts tend to be variable in size

References
ii. Donnelly LF. Fundamental of Pediatric Radiology. 2001 WB Saunders Company

40. With regard to air leak phenomenon in the neonate, which statement is true?

A. Air leak often enters a pulmonary vein.
B. Air leak in pulmonary interstitial emphysema can mimic a congenital pulmonary airway malformation.
C. Air leak occurs as a result of intubation and mechanical ventilation but not in the presence of continuous positive airway pressure (CPAP).
D. Pulmonary interstitial emphysema will not cause mediastinal shift.

Correct answer: B

Rationale
A: Air leak rarely enters the pulmonary vein. When this occurs, it may be fatal. B: Air leak in pulmonary interstitial emphysema may persist as a large cystic lesion that may mimic a congenital pulmonary airway malformation. C. Air leak often occurs as a result of intubation and mechanical ventilation but can also ensue with CPAP. D. When pulmonary interstitial emphysema is unilateral, it may cause mediastinal shift.

References
41. Which of the following diffuse lung disorders is not associated with respiratory distress in term or near-term neonates?
   A. Alveolar capillary dysplasia
   B. Surfactant protein B (SP-B) gene mutations
   C. ATP binding cassette A3 (ABCA3) gene mutations
   D. Neuroendocrine cell hyperplasia of infancy (NEHI)
   E. Primary ciliary dyskinesia

Correct Answer: D

Rationale
Answer is D. NEHI typically presents with persistent tachypnea, hypoxemia and slow weight gain in infants after the neonatal period. Alveolar capillary dysplasia usually presents within hours or days of birth with respiratory failure and severe persistent pulmonary hypertension due to markedly impaired alveolar gas exchange from pulmonary lobular maldevelopment. Neonatal respiratory distress rapidly progressing to respiratory failure can occur in term or near-term infants with biallelic autosomal recessive SP-B or ABCA3 gene mutations associated with deficient surfactant production. A majority of patients with primary ciliary dyskinesia experience neonatal respiratory distress related to impaired clearance of fetal lung fluid, but this is often misattributed to transient tachypnea of the newborn or neonatal pneumonia.

Reference

42. Which of the following statements about alveolar growth abnormalities is false?
   A. Most common cause of chronic diffuse neonatal lung disease
   B. Characterized by reduced alveolar size
   C. Often accompanied by pulmonary interstitial glycogenosis
   D. Associated with congenital heart disease and trisomy 21
   E. Decreased lung attenuation on CT correlates with clinical severity

Correct Answer: B

Rationale
Answer is B. Alveolar growth abnormalities are characterized by increased alveolar size and reduced alveolar number. This results in diminished alveolar septal surface area and vascularization, and the corresponding decreased lung parenchymal attenuation on CT correlates with clinical severity. Alveolar growth abnormalities are often accompanied by patchy pulmonary interstitial glycogenosis or pulmonary arterial hypertensive changes. Alveolar growth abnormalities are the most common cause of chronic diffuse neonatal lung disease, and include disorders such as bronchopulmonary dysplasia, Wilson-Mikity syndrome, and pulmonary hypoplasia. Alveolar growth abnormalities can also be associated with congenital heart disease and certain genetic disorders, such as trisomy 21 or filamin A gene mutations.

Reference
Neonatal Pulmonary Hypoplasia/Pulmonary Hypertension Therapeutic Options
Richard b. Parad, MD, MPH

43. Development of Pulmonary Hypoplasia is thought to most commonly occur during which of the following phases of lung development:
   A. Embryonic
   B. Pseudoglandular
   C. Canalicular
   D. Saccular
   E. Alveolar

Correct Answer: B

Reference

44. Which of the following is not associated with the development of pulmonary hypoplasia?
   A. Oligohydramnios
   B. Tracheal occlusion
   C. Interruption of vascular growth
   D. Space occupying lesions in the chest
   E. Diaphragmatic hernia

Correct Answer: B

Reference

45. The most sensitive measure of pulmonary hypoplasia of the newborn after delivery is:
   A. Chest radiography
   B. Chest MRI
   C. Lung wt/Body wt ratio
   D. pCO2
   E. Chest ultrasound

Correct Answer: C

Reference