Which of the following tumors is not known to be associated with the DICER1/PPB familial tumor predisposition syndrome in childhood or adolescence?

A. Pineoblastoma
B. Hepatoblastoma
C. Cystic nephroma
D. Ovarian Sertoli-Leydig tumor
E. Cervical embryonal rhabdomyosarcoma

- Correct Answer is B. Hepatoblastoma

- Characteristic tumors of the DICER1 familial tumor predisposition syndrome in childhood include pleuropulmonary blastoma, ovarian sex cord stromal tumors (especially Sertoli-Leydig cell tumor), differentiated thyroid carcinoma, cystic nephroma, cervical embryonal rhabdomyosarcoma, pituitary blastoma, pineoblastoma, nasal chondromesenchymal hamartoma, and ciliary body medulloepithelioma.

- This syndrome is not known to be associated with a predisposition to hepatoblastoma.

References:


Concerning clinical imaging of CF lung disease, which one of the following is correct?

A. Currently, CT should be considered the mainstay of routine sequential imaging in CF.
B. Persistent peribronchial thickening noted on CXR should be recognized as potentially the first manifestation of CF.
C. To date there is no evidence that MRI has a significant role in routine imaging of CF lung disease.
D. Recent research suggests that CT radiation dose will prevent the use of CT for routine sequential imaging in CF in the foreseeable future.
E. To answer the question, “What is acutely wrong?”, CXR is usually too nonspecific to prevent getting a CT.

Option A is not correct. The radiation dose and cost of CT currently preclude using CT as the routine standard imaging for sequential assessment of CF. However if imaging can be limited to specific areas, usually to evaluate specific issues such as progression of bronchiectasis, CT dose can be dramatically reduced.

Option B is correct. Commonly the first imaging manifestation of CF lung disease is diffuse persistent peribronchial thickening seen on CXR. A recent report on the use of MRI for CF lung disease reports that focal areas of air trapping, possibly recognized as focal areas of oligemia, may be detected before CXR findings are noted.

Option C is not correct. A recent report on the use of MRI (mentioned above), advocates the use of MRI as the routine imaging for CF lung disease.

Option D is not correct. A presentation at RSNA 2013 (from a group at the MGH) reports a CT protocol with a total dose equal to that of 10 PA CXR.

Option E is not correct. Acute deterioration is often related to issues such as bacterial pneumonia, pulmonary hemorrhage and pneumothorax. All may be readily apparent on CXR. More subtle or subacute deterioration may be seen with non-tuberculous mycobacteria, ABPA and increasing bronchiectasis, which although possibly detected by CXR, often require CT for definitive diagnosis and assessment.

REFERENCES

Spontaneous or idiopathic pulmonary embolism (PE) in children is rare compared to adults. Multiple co-existing risk factors are associated with PE in children. Which of the following is the most important risk factor for PE in children?

A. Indwelling central venous lines
B. Obesity
C. Malignancy
D. Sickle Cell Disease
E. Estrogen use
Answer is A. Indwelling central venous lines

Option A is correct. Central venous lines (CVL) are considered to be the most important risk factor for PE in children. The increased thrombotic events associated with CVL are thought to be due to altered regional blood flow, vessel wall injury and predisposition to infection. In the past decade, there has been an increasing use of CVL in children, mostly for chemotherapy, parenteral nutrition and drug administration. Several studies have shown that neonates are more prone (80-90%) to a venous thromboembolic disease compared to older children (30%-60%). Most CVL are placed in the upper extremity and may account for increased rates of PE associated with upper extremity deep venous thrombosis (DVT) in children (20%) compared to adults where a higher association of lower extremity DVT (95%) is found.

Option B is not correct. Obesity is a risk factor, but is not shown to be a major contributing risk factor in children compared to adults.

Option C is not correct. Malignancy confers a multifactorial risk for PE in children due to inherent coagulation abnormality, medications such as L-asparaginase for treating hematological malignancy and placement of CVL. However, it has been observed that the independent use of CVL for any clinical indication is more commonly associated with PE in children.

Option D is not correct. Sickle cell disease is known to cause a four-fold increase in the risk for PE in adults, but such an association has not been proven in children. It should be noted that pulmonary infarcts in sickle cell disease are due to fat embolism from bone infarction rather than venous thromboembolic disease.

Option E is not correct. Estrogen use causes a two to five fold increased risk of DVT and PE, but compared to CVL, it is relatively less common.

REFERENCE:

Concerning the mosaic attenuation pattern shown in the image

A. Areas of low attenuation are under ventilated with air trapping
B. Shunting of blood to the abnormal area results in over perfusion
C. Pulmonary embolism is the most common cause of the lucent areas
D. Increased attenuation corresponds to the abnormal areas.
E. Inspiratory scans differentiate air trapping from primary vascular abnormality

Answer = A, Areas of low attenuation are under ventilated with air trapping

Option A is correct. The lucent (low attenuation) areas are abnormal. These represent areas of air trapping. There is reflex vasoconstriction in these areas due to decreased gas exchange and blood is shunted away to the normally ventilated areas which have increased attenuation due to increased blood flow. Mosaic attenuation is seen in small airways disease.

Option B is not correct: Blood is shunted away from the abnormal areas. As there is decreased gas exchange due to air trapping, blood is shunted to the normal areas.

Option C is not correct: Mosaic attenuation is almost always due to small airways disease as pulmonary embolism is rare in children.

Option D is not correct: Areas of increased attenuation usually represent normally ventilated lungs and have relatively increased perfusion due to reflex vasoconstriction in the areas of air trapping.

Option E is not correct: Expiratory scans help to differentiate air trapping from other causes of mosaic attenuation. On expiration, the normal lung demonstrates an increase in attenuation, whereas the areas of air trapping do not demonstrate this change.
In performing dynamic pulmonary computed tomography, which of the following is a constant for all patient examinations?

A. kVp
B. mA
C. Gantry rotation speed
D. Continued PEEP
E. Z-axis length

Answer = A. kVp

The mA for each patient is individually determined for each patient using a weight based formula.

The gantry rotation speed is adjusted for each patient based upon the patient respiratory rate.

PEEP is turned off since the positive pressure will mask malacia.

The z-axis length is adjusted for each patient based on the size of the patient.

80 kVp is used for all patients since the structures to be visualized have high intrinsic contrast.

References:


When reducing the high voltage of the CT scanner in an effort to improve image quality and reduce the radiation dose to pediatric patients, for each type of clinical examination one can ignore the effect on:

A. Contrast.
B. Noise.
C. Sharpness.
D. Artifacts.
E. Scanning speed.

Answer is C, Sharpness

Option A is not correct because the contrast in the image is affected by the change in the high-voltage. Changes in high-voltage necessary to manage patient dose may have an adverse effect upon contrast.

Option B is not correct because the noise in the image is affected by the change in patient dose. Excessive reductions of patient dose may result in quantum model or noise in the image that is unacceptable.

Option C is correct. Changing the high-voltage or patient dose has no effect on the sharpness of the CT image.

Option D is not correct because reduced values of high-voltage in an effort to reduce the patient dose may introduce additional artifacts into the image.

Option E is not correct because reductions in high-voltage in an effort to reduce patient dose will increase the scan time to complete the entire exam. This may introduce artifacts or other issues in the images due to patient motion.

REFERENCES:


A 12-year-old girl post bone marrow transplant for immunodeficiency presents with inexorable breathlessness unresponsive to therapy.
A 12-year-old girl post bone marrow transplant for immunodeficiency presents with inexorable breathlessness unresponsive to therapy.

What is most likely diagnosis based on the HRCT?

A. Pulmonary interstitial glycogenolysis (PIE)
B. Neuroendocrine Hyperplasia of Infancy (NEHI)
C. Nonspecific interstitial pneumonitis (NSIP)
D. Pleuroparenchymal fibroelastosis (PPFE)
E. Pulmonary alveolar proteinosis (PAP)

Answer is D. Pleuroparenchymal fibroelastosis (PPFE)

PPFE (pleuroparenchymal fibroelastosis) occurred in this patient after bone marrow transplantation. This condition was previously reported as an idiopathic condition. This patient presented clinically with pneumothorax and sub pleural fibrosis on high-resolution computed tomography. In addition to the expected obliterative bronchiolitis, histopathology showed coexistent sub pleural changes. Options A, B, C and E are not correct although there are features similar to PPFE.

REFERENCES:


With MDCT as the reference standard, what is the smallest size of pulmonary nodule that can be detected with MRI with fast imaging sequences without contrast administration in the pediatric population?

A. 30 mm  
B. 10 mm  
C. 5 mm  
D. 3 mm

Answer is C, 5 mm

The recent prospective study consisted of 71 children who underwent paired thoracic MRI with fast imaging sequences and MDCT with intravenous contrast demonstrated that MRI with fast imaging sequences without contrast is comparable to contrast-enhanced MDCT for detecting pulmonary nodules larger than 3 mm (A, B, D are not correct).

Which of the following is true regarding pleuropulmonary blastoma (PPB) and DICER1 gene mutations?

A. Almost all PPBs are thought to be attributable to DICER1 mutation
B. DICER1 mutations predisposing to PPB are typically spontaneous rather than inherited, so that family members can usually be reassured that they are not at higher risk of tumors
C. Most individuals with DICER1 mutations develop PPB
D. Chest imaging screening for cystic PPB should be considered in infant carriers of DICER1 mutations

REFERENCE:

Correct Answer is D. Chest imaging screening for cystic PPB should be considered in infant carriers of DICER1 mutations

Tumors in the DICER1 familial tumor predisposition syndrome tend to follow a two-hit model of tumorigenesis, in which an inherited heterozygous loss of function germline DICER1 mutation is followed by a somatic “hot spot” DICER1 mutation leading to neoplastic transformation. About 70% of individuals with PPB have heterozygous loss of function germline DICER1 mutations, and DICER1 mutation testing should be conducted in the proband with PPB and family members for carrier identification and genetic counseling. The penetrance of PPB in DICER1 germline mutation carriers is estimated at 10-15%. Although the penetrance is low, chest imaging screening for cystic PPB should be considered in infant carriers of DICER1 mutations due to the risk of progression of undetected unresected cystic PPB to aggressive, high-grade, solid PPB.

Images of the trachea during inspiration and expiration phases of a dynamic pulmonary computed tomography examination in a 2 month old infant are shown. A 43% reduction in trachea cross-sectional imaging during expiration was measured. The trachea should be reported as:

A. Normal
B. Tracheal atresia
C. Tracheomalacia
D. Trachea stenosis
E. Extrinsic compression

Answer = C. Tracheomalacia

The cross sectional area reduction is too great to be considered normal. A reduction of trachea cross sectional area during expiration of greater than 28% is consistent with tracheomalacia during a normal breathing cycle. Greater than 50% reduction in cross sectional area is used as a criterion for tracheomalacia in older patients performing a forced expiration. The correct answer is C. Tracheomalacia.

The reduction in cross sectional area is not fixed as would be expected in trachea stenosis.

No atresia is present.

No extrinsic compression is demonstrated.

References:
A 3-month-old full term old girl with nystagmus and deteriorating respiratory function. HRCT of lung parenchyma is most likely to represent which of the following?

A. BPD  
B. Meconium aspiration  
C. Congenital lobar over inflation  
D. Congenital surfactant deficiency  
E. Filamin protein deficiency related infantile multilobar emphysema

Answer is E. Filamin protein deficiency related infantile multilobar emphysema.
REFERENCES:


2. Imaging of ChILD; Guillerman RP. Pediatric allergy immunology and Pulmonology vol 23; number 1 2010.

Which of the following statements comparing PE in children and adults is NOT true?

A. Wells and Geneva clinical probability scores are not validated in children.

B. PE is an uncommon cardiovascular disease in children.

C. PE in children is commonly unprovoked or idiopathic in nature.

D. CT pulmonary angiography (CTPA) is the most commonly used diagnostic test in evaluating PE.

E. Upper extremity DVT associated with PE is more commonly seen in children than adults.

Answer is C. PE in children is commonly unprovoked or idiopathic in nature.

Option A is not correct. Clinical probability scores are useful to assess pre-test probability of PE in adults, however they have not been validated in children. Data on PE in children suggests a high association in the presence of multiple risk factors.

Option B is not correct. PE is rare in children compared to adults where it is the third most common cause of acute cardiovascular disease.

Option C is correct. Idiopathic or unprovoked PE is more common in adults with an overall incidence of 30% compared to 0-4% in children. Most cases of PE in children are associated with predisposing risk factors.

Option D is not correct. Imaging studies for evaluation of PE in children include ventilation perfusion scintigraphy (VQ scan), CTPA and conventional pulmonary angiography. In the past decade with the wide spread availability and technological advancement of CT, CTPA has become the frontline imaging test and standard of reference for evaluating PE. Improvement in CT technology not only allows for routine visualization of pulmonary arteries to the sub-segmental level but also helps with alternative diagnosis. However, the major concern of CTPA is ionizing radiation. It has been shown that CTPA has a relatively higher radiation dose than VQ scan and should be judiciously used in children.

Option E is not correct. In adults, approximately 95% of PE is attributed to lower extremity DVT, whereas in children upper extremity DVT is seen in up to 20% of cases. This is thought to be due to the increased use of upper extremity CVL in children who have relatively smaller caliber vessels.

REFERENCES:


With respect to the reduction of mAs when developing thoracic CT technique factors for a 1-year-old patient:

A. 1-year-old (1Y) dose = adult dose (AD) if adult mAs unchanged
B. 1Y dose = ½ of AD if adult mAs divided by 2
C. 1Y dose = AD dose if adult mAs divided by 3
D. 1Y dose = ½ of AD dose if adult mAs divided by 4
E. 1Y dose = ½ of AD dose does not provide clinically useful images

Answer = D

Option A is not correct because if the adult mAs is unchanged, the 1Y dose will be at least double the adult patient dose.

Option B is not correct because if the adult mAs is divided by 2, the 1Y dose will = the adult patient dose.

Option C is not correct because if the adult mAs is divided by 3, the 1Y dose will be 2/3 of the adult patient dose.

Option D is correct. One quarter of the mAs of an adult technique delivers half of the adult dose to a 1Y patient.

Option E is not correct because the first reference determined that pediatric radiologists found 1Y CT images with dose ½ of adult doses to be clinically acceptable.

REFERENCES:


HRCT technique requires which of the following?

A. Scanning during maximum inspiration
B. Thin images with high-resolution reconstruction
C. Thick reconstructions for mediastinal evaluation
D. Step-and-shoot axial scanning
E. Rapid bolus intravenous contrast injection
Answer = B, Thin images with high-resolution reconstruction

Option A is not correct: HRCT images can be obtained in inspiration, expiration or quiet respiration. In some selective cases, both inspiratory and expiratory scans are obtained to diagnose air trapping.

Option B is correct: HRCT involves obtaining thin images and reconstructing them using high resolution algorithm to give sharper images of the lungs. Traditionally 1mm axial images were obtained with a gap of 10mm. However with the multidetector CTs, a rapid acquisition of volumetric images of the entire lung is possible and the images can be reconstructed using the HRCT algorithm.

Option C is not correct: The reconstruction algorithm used in HRCT makes the images of low contrast structures like the mediastinum suboptimal due to increase in noise. HRCT reconstructions are for evaluating high contrast structures such as the lungs.

Option D is not correct: HRCT images can be obtained by applying the reconstruction algorithm to volumetric images obtained by MDCT. Some studies have shown that motion artifacts are less with this technique compared to axial acquisition of the images.

Option E is not correct: Intravenous contrast is not essential to obtain HRCT images.

Concerning the Brasfield and Wisconsin CXR based scoring systems, which one of the following is correct?

A. PFT (pulmonary function tests) are more sensitive in detecting differences in groups of CF patients than are CXR based scoring systems.

B. The relative simplicity of the Brasfield system compromises its reliability.

C. The complexity of the Wisconsin system compromises its reliability.

D. The Brasfield and Wisconsin systems perform equally well in terms of intra-rater and inter-rater reliability and in comparison to PFT.

E. Scoring of CT outperforms either CXR based system in predicting future lung disease severity.

References:
REFERENCES


MRI with fast imaging sequences without contrast administration is comparable to contrast-enhanced MDCT for detecting thoracic abnormalities in pediatric patients EXCEPT:

A. Pneumonia
B. Congenital foregut duplication cyst
C. Pleural effusion
D. Interstitial lung disease

Answer is D, Interstitial Lung Disease

The recent prospective study consisted of 71 children who underwent paired thoracic MRI with fast imaging sequences and MDCT with intravenous contrast demonstrated that MRI with fast imaging sequences without contrast is comparable to contrast-enhanced MDCT for detecting a variety of thoracic abnormalities including infections, congenital foregut duplication cysts, and pleural effusions (A, B, C are not correct).

However, MRI is less sensitive than MDCT for detecting underlying mild interstitial lung disease and small pulmonary nodules (D is correct).

REFERENCES:
