

SPR 2014 Thoracic Imaging Session
May 17, 2014
SAM Questionnaire

Thoracic Manifestations of Systemic Disease in Children: Imaging Clues to Diagnosis

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1. **Systemic conditions that are typically associated with cystic lung changes include all of the following EXCEPT.**
 - A. Down syndrome
 - B. Wegener's granulomatosis
 - C. Langerhans cell histiocytosis
 - D. Sickle cell disease
 - E. Proteus syndrome

Correct Answer: D

References

1. Biko DM, Schwartz M, Anupindi SA, Altes TA. Subpleural lung cysts in Down syndrome: prevalence and association with coexisting diagnoses. *Pediatric Radiology*. 2008; 38(3):280-4.
2. Martinez F, Chung JH, Digumarthy SR, et al. Common and uncommon manifestations of Wegener granulomatosis at chest CT: radiologic-pathologic correlation. *Radiographics* . 2012; 32(1):51-69.
3. Bano S, Chaudhary V, Narula MK et al. Pulmonary Langerhans cell histiocytosis in children: A spectrum of radiologic findings. *European Journal of Radiology*. 2014; 83(1):47-56.
4. Leong CS, Stark P. Thoracic Manifestations of Sickle Cell Disease. *J Thorac Imaging*. 1998; 13(2):128-34.
5. Newman B, Urbach A, Orenstein D, Dickman PS. The Proteus syndrome: Emphasis on the Pulmonary Manifestations. *Pediatric Radiology*. 1994; 24: 189-193.

Rationales

- A. is **not** correct. Approximately 1/3 of Down syndrome patients exhibit small subpleural and perifissural cysts on chest CT imaging. The etiology is uncertain but is thought to be related to pulmonary hypoplasia and an alveolar growth disorder.
- B. is **not** correct. Wegener's granulomatosis is a vasculitic condition, most common in teenage girls, affecting small to medium-size vessels particularly in the upper respiratory tract, lungs, & kidneys. Imaging findings in the lungs are characterized by multifocal parenchymal airspace and ground glass opacities and/or nodules with cavitation/cystic change or surrounding ground glass (hemorrhage).
- C. is **not** correct. Langerhans cell histiocytosis with systemic disease most commonly affects males between 1-3yrs old with lung involvement in about 30%. Chest radiographic findings include reticulonodular interstitial opacities progressing to a cystic pattern. May develop pneumomediastinum or pneumothorax. CT changes consist of multiple small nodules that become cystic and may progress to fibrosis, adenopathy is common.
- D. is **correct**. Sickle cell acute chest syndrome is characterized by chest pain with focal pulmonary infiltrate/s and pleural effusion. While it is thought to be precipitated by microvascular occlusion, the radiographic changes may be due to superimposed infection,

infarction or atelectasis. Other imaging findings may include cardiomegaly, congestion and bone changes secondary to infarction in ribs, humeri and vertebral bodies. In theory pulmonary cystic change might develop in association with infection or infarction but this would be an atypical feature of sickle cell disease in the chest.

- E. is **not** correct. Proteus syndrome is a hamartomatous disorder with a large variety of features that include hemihyperplasia, macrodactyly, subcutaneous masses, vascular abnormalities, palmar and plantar fibrous overgrowth, exostoses, epidermal nevi and scoliosis. Pulmonary cystic changes can be very prominent and even life threatening and are associated with panlobar emphysema.

2. Which of the following statements regarding portopulmonary syndrome is CORRECT?

- A. Progressive hypoxemia and pulmonary hypertension are uncommon presenting features.
B. Both polysplenia and asplenia are associated.
C. “Hepatic factor” does not reach the pulmonary circulation with resultant diffuse pulmonary vasodilatation.
D. It is usually associated with severe underlying liver dysfunction.
E. Liver transplantation is the most effective treatment.

Correct Answer: C

References:

1. Kinane TB, Westra SJ. Case records of the Massachusetts General Hospital. Weekly clinicopathological exercises. Case 31-2004. A four-year-old boy with hypoxemia. *New Engl J Med.* 2004; 351:1667-1675.
2. Newman B, Feinstein JA, Cohen RA, Feingold B, Kreutzer J, Patel H, Chan FP. Congenital extrahepatic portosystemic shunts associated with heterotaxy and polysplenia. *Pediatr Radiol.* 2010; 40(7):1222-30.
3. Gupta NA, Abramowsky C, Pillen T, *et al.* Pediatric hepatopulmonary syndrome is seen with polysplenia/interrupted inferior vena cava and without cirrhosis. *Liver Transplant.* 2007; 13:680-686.

Rationales

- A. is **not** correct. The typical clinical presentation is progressive hypoxemia due to diffuse intrapulmonary arteriovenous shunting. Pulmonary hypertension is also usually present.
- B. is **not** correct. Congenital extrahepatic portosystemic shunts that produce portopulmonary syndrome are associated with heterotaxy and polysplenia but not asplenia.
- C. is **correct**. Although it is uncertain what the “hepatic factor” actually is, it appears that mesenteric venous blood must pass through the liver before reaching the pulmonary circulation in order to produce a normal balance of vasodilation/vasoconstriction. When the circulation bypasses the liver due to intra or extrahepatic portosystemic shunting or surgical exclusion of hepatic blood flow from the lungs, the result is diffuse peripheral pulmonary vasodilation and arteriovenous shunting.
- D. is **not** correct. While hepatopulmonary syndrome produces a similar end result in the lungs and is associated with severe hepatic dysfunction, liver function is typically normal in portopulmonary syndrome with congenital portosystemic connections as the usual underlying etiology.
- E. is **not** correct. Liver transplantation is the treatment of choice when portopulmonary syndrome is accompanied by an absent portal vein. However portopulmonary syndrome may persist or recur after liver transplantation if a large portosystemic shunt is not recognized

and ligated. When portal and hepatic veins are intact, the treatment of choice is closure of the portosystemic shunt in order to reroute blood flow through the liver. Completion Fontan surgery with routing of hepatic blood flow to the lungs is the management used in the case of single ventricle superior cavopulmonary shunts.

- 3. Long segment tracheal stenosis with complete tracheal rings is associated with:**
- A. Tracheoesophageal fistula
 - B. Granulomatosis with polyangiitis (GPA)
 - C. Tetralogy of Fallot
 - D. Vascular ring
 - E. Congenital diaphragmatic hernia

Correct Answer: C

References

1. Kazim R, Berdon WE, Montoya CH, Quaeghebeur JM, Sun LS. Tracheobronchial anomalies in children with congenital cardiac disease. *J Cardiothorac Vasc Anesth.* 1998 Oct;12(5):553-5.
2. Kazim R, Quaeghebeur JM, Sun LS. The association of tracheal anomalies and tetralogy of Fallot. *J Cardiothorac Vasc Anesth.* 1996 Aug;10(5):589-92.
3. Starc MT, Berdon WE, Starc TJ. Undiagnosed primary tracheal stenosis in tetralogy of Fallot: complete rings with a low carina. *Pediatr Radiol.* 2013 Dec 27

Rationales

- A. is **not** correct. Tracheoesophageal fistula is usually associated with severe tracheomalacia in the area of the fistula after its repair. The malacia tends to be severe enough to cause complete collapse of the tracheal lumen (especially during periods of agitation and/or coughing) that leads to profound oxyhemoglobin desaturation (the so-called “TEF death spells”). The severity of the spells diminishes gradually during the first several years of life. However, the affected patients tend to have symptoms of “chronic bronchitis” due to difficulty in clearing secretions.
- B. is **not** correct. Granulomatosis with polyangiitis (GPA) (formerly known as Wegener’s granulomatosis) is one of the vasculitides that affect small and medium sized vessels in multiple organs. One of its serious complications is rapidly progressing renal failure. In the lung it can cause pulmonary hemorrhage (that occasionally may be the presenting symptom). The condition is also associated with subglottic stenosis but not tracheal stenosis.
- C. is **correct**. Of the conditions mentioned above, Tetralogy of Fallot, is most often associated with long segment tracheal stenosis with complete tracheal rings, although the association is less common than that of pulmonary sling. The carina tends to be in a lower position than normal and the main stem bronchii tend to be attached to the trachea in an almost 90° angle giving a characteristic shape of an inverted capital T. Affected infants tend to have biphasic (inspiratory and expiratory) wheeze or stridor and they are often misdiagnosed as “asthmatics”..Airway abnormality should be considered and evaluated in symptomatic patients.
- D. is **not** correct. Vascular rings tend to cause localized compression of the trachea resulting in narrowing of the tracheal lumen and they may cause localized tracheomalacia. However, the affected segment tends to be short and the tracheal rings are C-shaped and not complete.
- E. is **not** correct. Congenital diaphragmatic hernia (CDH) is associated with lung hypoplasia on the affected side and in severe cases with at least microscopic underdevelopment of the

contralateral lung (e.g. fewer generations of bronchii, incomplete branching of the bronchioles and/or of the pulmonary vessels as well as underdevelopment of the alveoli.

Imaging Evaluation of Pediatric Airway and Lung Neoplasms: What Does the Clinician Need to Know?

Paul Thacker, MD

4. In comparison of the WHO, RECIST 1.0, and RECIST 1.1 criteria, which of the following is the best answer?
- A. All three criteria define a measurable lesion as a minimum size of 10 millimeters.
 - B. RECIST 1.1 uses the sum of products of diameters as the method for lesion measurement.
 - C. RECIST 1.0 defines progressive disease as $\geq 25\%$ growth in the single longest diameter.
 - D. When measuring lymph nodes to assess for pathologic enlargement, the shortest diameter is measured according to RECIST 1.1 criteria

Correct Answer: D

References

1. Tirkes T, Hollar MA, Tann M, et al. Response criteria in oncologic imaging: review of traditional and new criteria. *Radiographics* 2013; 33: 1323-1341.
2. Chalian H, Tore HG, Horowitz JM, et al. Radiologic assessment of response to therapy: comparison of RECIST versions 1.1 and 1.0. *Radiographics* 2011; 31: 2093-2105.
3. Van Meerten EL, Gelderblom H, and Bloem JL. RECIST revised: implications for the radiologist. A review article on the modified RECIST guideline. *Eur Radiol* 2010; 20: 1456-1467.
4. McHugh K and Kao S. Response evaluation criteria in solid tumours (RECIST): problems and need for modifications in paediatric oncology? *British Journal of Radiology* 2003; 76: 433-436.

Rationales

Answer is D.

The question stem asks to compare the WHO, RECIST 1.0, and the modified RECIST criteria (RECIST 1.1).

Option A is not correct. The WHO criteria has no minimum lesion size. Thus, any lesion is measurable and should be measured in two dimensions.

Option B is not correct. RECIST 1.1 uses the longest diameter as the method for target lesion measurement, with the exception of lymph nodes which are measured in the shortest diameter. Of the three criteria, the only one which uses the sum of products of diameters (i.e. longest overall tumor diameter and longest diameter perpendicular to the longest overall diameter) is the WHO criteria.

Option C is not correct. RECIST 1.0 defines progressive disease as a 20% increase in the single longest diameter or new lesions. WHO criteria defines progressive disease as $\geq 25\%$ increase in the sum of the products of diameters.

Option D is correct. Lymph nodes are assessed for pathologic enlargement using the shortest diameter with target lesions defined as a short axis diameter ≥ 15 mm, nontarget lesions = 10-15 mm, and nonpathologic lesions < 10 mm.

5. Figure 1 shows an image from a chest CT (soft tissue window) of a 4-year-old male with a right lower hemithorax pleuropulmonary blastoma. Which one of the following statements is most correct?



- A. Pleuropulmonary blastomas represent the most common malignant lung parenchymal tumor of childhood with an annual incidence of 1:100,000.
- B. There are 4 types of pleuropulmonary blastomas defined based on the ratio of solid and cystic components.
- C. Pleuropulmonary blastomas have been reported to arise within pulmonary airway malformations, sequestrations, and bronchogenic cysts.
- D. The prognosis for patients with pleuropulmonary blastoma is poor with a long-term survival of < 25%.

Correct Answer: C

References

1. Cleveland RH, ed. Imaging in Pediatric Pulmonology. New York: Springer, 2012; 270-271.
2. Federici S, Domenichelli V, Tani G, et al. Pleuropulmonary blastoma in congenital cystic adenomatoid malformation: report of a case. Eur J Pediatr Surg 2001; 11(3): 196-199.
3. Naffaa LN, Donnelly LF. Imaging findings in pleuropulmonary blastoma. Pediatr Radiol 2005; 35: 387-391.

Rationale

Answer C is correct.

Option A is not correct. Although pleuropulmonary blastomas are thought to be the most common malignant parenchymal tumor of childhood, the actual incidence is unknown.

Option B is not correct. There are three types of pleuropulmonary blastomas. Type I is entirely cystic and is difficult to distinguish from type I congenital pulmonary airway malformations. Type II contains both cystic and solid components. Type III is predominantly solid.

Option C is correct. Pleuropulmonary blastomas have been reported to arise in multiple congenital lung anomalies including bronchogenic cysts, sequestrations, and congenital pulmonary airway malformations.

Option D is not correct. Prognosis of pleuropulmonary blastomas is dependent on type with type I having an 83% long-term survival. Up to 30% of patients with types II and III pleuropulmonary blastoma have metastasis to bone, liver and brain with an overall long-term survival rate of 42%.

Pediatric Mediastinum: Clinical Correlation and Practical Imaging Assessment

Maryam Ghadimi Mahani, MD

6. What is the radiological sign in this image and what does it refer to?

- A. "Wave sign" in a normal thymus
- B. "Hilum over lay sign" in an anterior mediastinal mass
- C. "Wave sign" in an anterior mediastinal mass
- D. "Hilum over sign" in middle mediastinal mass



Correct Answer: B

“The hilum overlay sign is present when the normal hilar structures project through a mass, such that the mass can be understood as being either anterior or posterior to the hilum.”

References

1. Whitten CR, Khan S, Munneke GJ, Grubnic S. A Diagnostic Approach to Mediastinal Abnormalities. *Radiographics*. 2007 May-Jun; 27(3):657-71.

Wave sign is the indentations produced by anterior ribs on the soft thymus, another normal pattern. The lobular border of this mass is not the wave sign.

2. Shashi H. Ranganath¹, Edward Y. Lee¹, Ricardo Restrepo² and Ronald L. Eisenberg. Mediastinal Masses in Children. *American Journal of Roentgenology*. 2012;198: W197-W216.10.2214/AJR.11.7027
7. **In evaluation of the mediastinal masses on CT which is more helpful in differentiation between a normal thymus and abnormal thymus or a mediastinal mass?**
 - A. Size
 - B. Homogenous versus inhomogeneous parenchyma
 - C. Mass effect over adjacent structures
 - D. Normal thymus gland does not have retrocaval extension
 - E. Normal thymus does not extend to lower neck

Correct Answer: C

References

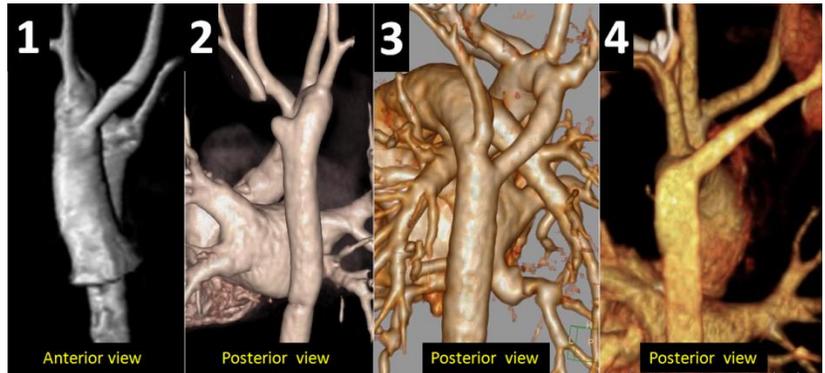
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2. Frush DP. Imaging evaluation of the thymus and thymic disorders in children. In: Lucaya J, Strife JL, eds. *Pediatric chest imaging*. Berlin, Germany: Springer-Verlag, 2001:187-208
3. Costa NS, Laor T, Donnelly LF. Superior cervical extension of the thymus: a normal finding that should not be mistaken for a mass. *Radiology* 2010 Jul;256(1):238-42. doi: 10.1148/radiol.10091792. Epub 2010 May 26.

Pediatric Thoracic Vascular Imaging: How I Do It

Monica Epelman, MD

8. You are shown the following volume rendered 3D images. Which of the following examples constitute a vascular ring?

- A. 1, 2, and 3
- B. 2 and 3
- C. Only 3
- D. None of the above
- E. All of the above



Correct Answer: A

Rationale

Option A is correct. Complete vascular rings are composed often of atretic portions, which is problematic, as the atretic portion is not apparent with current imaging techniques. In these instances, there are three extremely useful secondary signs, all of which begin with the letter “D”, that should be considered [1-3] and that should raise suspicion of the presence of a vascular ring. These secondary signs include **d**iverticulum, **d**imple, and the course of the **d**escending aorta, all of which affect the opposite side of the aortic arch [1-3]. The presence of a **d**imple or a **d**iverticulum, that is larger in caliber than the aberrant subclavian artery, always implies the existence of a vascular ring. On the other hand, the simple aberrant subclavian artery is uniform in caliber. The rationale for this is that during fetal life the patent ductus arteriosus (PDA) connects to the descending aorta and carries more than 50% of the combined output of the two ventricles, while the aberrant subclavian artery only supplies the arm. Following ductal closure, the aberrant subclavian artery has a large proximal portion (diverticulum) and a smaller distal portion, although the flow in each portion is similar, the difference in caliber remains throughout life.

Image 1 reflects a right aortic arch with an aberrant left subclavian artery off a Kommerell **d**iverticulum, image 2 represents a double aortic arch with an atretic segment seen in the left arch following the take-off of the left subclavian artery. The atretic portion of the ligamentum arteriosum completes the ring, but is not visible with current imaging techniques. Its presence can be inferred by the existence of the **d**imple noted in the descending aorta on the opposite side of the arch. Image 3 depicts a right aortic arch with a left **d**escending aorta (right circumflex aortic arch). The **d**escending aorta in this case is opposite the side of the arch. There is, in addition, a large PDA, which completes the ring. Note the aberrant left subclavian artery originating from the PDA, the aberrant subclavia is smaller in caliber when compared to the PDA. Therefore, option A is correct, as images 1, 2 and 3 depict vascular rings. Image 4 reflects a left aortic arch with an aberrant right subclavian artery; the aberrant vessel is smooth in caliber throughout its course, no diverticulum is observed at its origin and there is no vascular ring in these cases.

Option B is not correct. Image 1, which reflects a right aortic arch with an aberrant left subclavian artery off a Kommerell diverticulum, also constitutes a vascular ring. Since it is not listed in this option, this option is incorrect.

Option C is not correct. Image 1, which reflects a right aortic arch with an aberrant left subclavian artery off a Kommerell diverticulum and image 2, which represents a double aortic arch with an atretic segment seen in the left arch following the take-off of the left subclavian artery also

constitutes a vascular ring, since these two options are not listed in this answer, this option is **incorrect**. Note the aortic dimple in the descending aorta on the opposite side of the arch in image B.

Option D is not correct. This option is incorrect as 1, 2 and 3 constitute examples of vascular rings.

Option E is not correct. This option is incorrect since 4 does not constitute an example of a vascular ring.

9. Which of the following findings is unexpected in pulmonary arterial hypertension and should be carefully ruled out before instituting treatment with pulmonary vasodilators, as if present and overlooked, it may result in fatal pulmonary edema?

- A. Pulmonary vein stenosis
- B. Peripheral tapering with pruning of the distal pulmonary arterial vasculature
- C. Right ventricular hypertrophy, with right ventricular and atrial dilatation
- D. Flattening or reversal of the interventricular septum
- E. Reflux of IV contrast into the inferior vena cava and hepatic veins

Correct Answer: A

Rationale

Option A is correct. In cases in which pulmonary hypertension is the result of downstream obstruction, such as pulmonary vein stenosis, pulmonary veno-occlusive disease, and left atrial hypertension, the use of pulmonary vasodilators prior to the relief of these obstructions may result in acute, life-threatening pulmonary edema. Therefore, all these conditions must be sought and carefully excluded.

Option B is not correct. The imaging findings of pulmonary hypertension on cross-sectional imaging include enlargement of the central pulmonary arteries, peripheral tapering with pruning of the distal pulmonary arterial vasculature, right ventricular hypertrophy, and right ventricular and atrial dilatation with flattening of the interventricular septum, posterior displacement of the left ventricle by the enlarged right cardiac chambers and reflux of IV contrast into the inferior vena cava and hepatic veins [4-6]. Peripheral tapering with pruning of the distal pulmonary arterial vasculature is an expected imaging finding in pulmonary arterial hypertension, which helps to characterize the condition, therefore this option is incorrect.

Option C is not correct. Right ventricular hypertrophy, with right ventricular and atrial dilatation are expected imaging findings in pulmonary hypertension.

Option D is not correct. Flattening or reversal of the interventricular septum is one of the expected findings in pulmonary arterial hypertension and it is regarded as one of the most valuable CT signs of right ventricular dysfunction.

Option E is not correct. Reflux of IV contrast into the inferior vena cava and hepatic veins is an expected imaging finding in the setting of pulmonary arterial hypertension and is reflective of high pressures within the right chambers.

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2. Weinberg PM, Whitehead KK (2010) Aortic Arch Anomalies. In: Fogel MA (ed) Principles and practice of cardiac magnetic resonance in congenital heart disease: Form, function and flow. Wiley-Blackwell, pp 183-208.

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4. Grosse C, Grosse A (2010) CT findings in diseases associated with pulmonary hypertension: a current review. Radiographics 30:1753-1777.
5. Frazier AA, Burke AP (2012) The imaging of pulmonary hypertension. Seminars in ultrasound, CT, and MR 33:535-551.
6. Friesen RH, Williams GD (2008) Anesthetic management of children with pulmonary arterial hypertension. Paediatric anaesthesia 18:208-216.

Pediatric Lung Transplantation: Clinical Perspectives and Imaging Assessment

Debra Boyer, MD and Edward Y. Lee, MD, MPH

- 10. What is the primary cause of death in lung transplant recipients greater than one year out from their transplant?**
- A. Ischemia-reperfusion injury
 - B. Post-transplant lymphoproliferative disease
 - C. CMV pneumonitis
 - D. Bronchiolitis obliterans syndrome (BOS)

Correct Answer: D

Rationale

Answer is D. Bronchiolitis obliterans syndrome occurs in approximately 50% of lung transplant recipients within five years post lung transplantation. This risk appears to be somewhat decreased in infants. Clinical symptoms consist most commonly of an insidious onset of dyspnea and hypoxia, however, occasionally the onset can be more rapid in evolution.

Option A is not correct. Ischemia-reperfusion injury occurs within the first 72 hours of lung transplantation and is related to the development of primary graft dysfunction.

Option B is not correct. Post-transplant lymphoproliferative disease (most often EBV-related) is the most common malignancy seen after lung transplantation. PTLD is more common in children than adults, however, as a cause of death PTLD is less common than BOS.

Option C is not correct. CMV disease can be a serious complication of lung transplantation and can contribute to significant morbidity and mortality. However, it is less common as a cause of death than BOS.

References

1. Sweet SC. Pediatric lung transplantation. Proc Am Thorac Soc. 2009; 6(1): 122-7.
 2. Solomon M, Grasemann H, Keshavjee S. Pediatric lung transplantation. Pediatr Clin North Am. 2010; 57(2): 375-91.
- 11. Predisposing factors to infection in the lung transplant recipient include all of the following EXCEPT:**
- A. Direct communication with the external environment
 - B. Immunosuppressed status
 - C. Abnormal mucociliary clearance
 - D. Bronchial anastomosis
 - E. Opioid therapy prior to lung transplant

Correct Answer: E

Rationale

E is the correct answer. While many lung transplant recipients receive opioids prior to their lung transplant for pain and/or dyspnea, these medications do not themselves increase the recipient's risk of developing an infection in the post lung transplant period.

Option A is not correct. As the lungs are the only transplanted organ with a direct connection to the external environment, they have a significantly increased risk of infection.

Option B is not correct. Due to the substantial risk of rejection in the lung transplant patients, they receive high doses of immunosuppressive medication both in the post-operative period and for the remainder of their lives. This significantly increases their risk of developing infectious complications.

Option C is not correct. There is significantly abnormal mucociliary clearance in the lung transplant patient in the post-operative period. This inability to properly clear secretions increases the risk of infections.

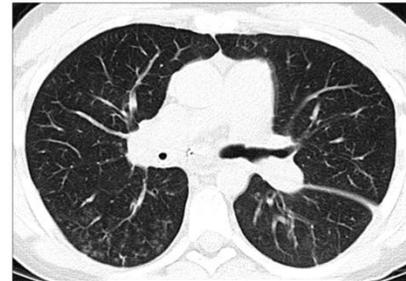
Option D is not correct. As the bronchial arteries are not reanastomosed at the time of the lung transplant, the mainstem bronchial anastomosis receive their blood supply through retrograde flow. This can lead to devitalized tissue, especially in the immediate post-operative period and predispose to airway infections, particularly fungal.

References

1. Ahmad S, Shlobin OA, Nathan SD. Pulmonary complications of lung transplantation. Chest. 2011; 139(2): 402-11. Sims KD, Blumberg EA. Common infections in the lung transplant recipient. Clin Chest Med. 2011; 32(2): 327-41.
2. Sims KD, Blumberg EA. Common Infections in the lung transplant recipient. Clin Chest Med. 2011; 32(2): 327-41.

12. 15-year-old girl, who underwent bilateral lung transplantation 2 years ago, presents with progressively worsening shortness of breath. Based on this axial CT image (Figure 1), what is the MOST likely etiology of the patient's respiratory symptom?

- A. Hyperacute rejection
- B. Fungal infection
- C. Large airway stricture
- D. Posttransplant lymphoproliferative disorder



Correct Answer: C

Rationales

Answer is C. The CT demonstrates markedly narrowed right main stem bronchus consistent with a large airway stricture. Left bronchus is normal in size.

Option A is not correct. Hyperacute rejection is due to recipient antibody response to donor vascular endothelium. It occurs minutes to hours after lung transplantation unlike in this patient who presented with progressively worsening shortness of breath. On imaging studies, hyperacute rejection is characterized by severe pulmonary edema and diffuse consolidation. Large airway is not typically involved.

Option B is not correct. Candida Albicans and Aspergillus are the two most common causes of fungal infection in pediatric patients with lung transplantation. Fungal infection in this patient population is characterized by consolidation, ground glass or cavitary opacities and ill-defined nodules. Large airway is not typically involved.

Option C is correct. Bronchial stricture, a potentially serious complication after lung transplantation, is due to a complication of surgical anastomotic healing or dehiscence. CT is currently the imaging modality of choice for non-invasively evaluating bronchial stenosis after lung transplantation in children. On CT, narrowing of the bronchus can be easily recognized.

Option D is not correct. Posttransplant lymphoproliferative disorder (PTLD) typically presents as solitary or multiple pulmonary nodules or masses in addition to often necrotic mediastinal lymphadenopathy approximately 1 year after the lung transplantation. Large airway involvement is rare.

References

1. Siegel MJ, Lee EY, Sweet SC, Hildebolt C. CT of posttransplantation lymphoproliferative disorder in pediatric recipients of lung allograft. *AJR Am J Roentgenol.* 2003; 181(4): 1125 – 1131.
2. Lee EY, Siegel MJ. MDCT of tracheobronchial narrowing in pediatric patients. *J Thorac Imaging.* 2007; 22(3): 300 – 309.

13. Which CT imaging finding is most sensitive for diagnosing bronchiolitis obliterans (BO) in pediatric patients with lung transplantation?

- A. Pulmonary edema
- B. Multiple pulmonary nodules
- C. Necrotic mediastinal lymphadenopathy
- D. Air trapping

Correct Answer: D

Rationale

Answer is D. Bronchiolitis obliterans (BO) is the major limitation for long-term survival and a frequent complication in pediatric patients with lung transplantation. On CT, BO is characterized by mosaic attenuation, bronchial dilatation, and bronchial wall thickening, and air trapping seen on expiratory CT images.

Option A is not correct. Pulmonary edema is typically seen in the setting of volume overload related to transfusion during lung transplant surgery or hyperacute rejection.

Option B is not correct. Multiple pulmonary nodules are typically seen in the setting of underlying infectious process in pediatric patients with lung transplant.

Option C is not correct. Necrotic mediastinal lymphadenopathy is seen in the setting of underlying infection or posttransplant lymphoproliferative disorder (PTLD).

Option D is correct. Air trapping on expiratory CT can help accurately diagnosing BO in pediatric lung transplant patients with 100% sensitivity, 71% specificity, 64% positive predictive value, and 100% negative predictive value.

Risks and Benefits of Pediatric Thoracic CT: Updated Information for Clinicians

Sjirk J. Westra, MD

14. For pediatric thoracic disease, CT has been shown to have the highest yield in:

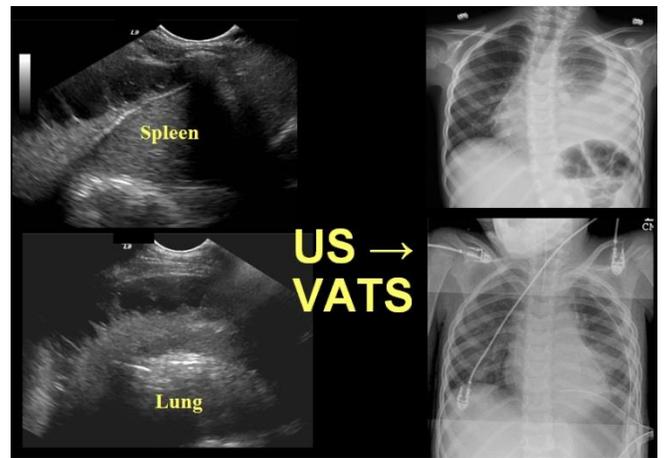
- A. Diffuse interstitial disease (e.g. NEHI)
- B. Localized lung disease (e.g. pneumonia)
- C. Pleural disease (e.g. empyema)
- D. Trauma (e.g. contusion, PNTX)
- E. Congenital malformations (e.g. pulmonary sequestration)

Correct Answer: E

Reference: Schneebaum Pediatrics 2009

15. 7-year-old previously healthy boy with pneumonia who continued to have fever despite 4 days of antibiotics. The following additional study is indicated:

- A. Frontal decubitus radiographs to demonstrate shifting of effusion
- B. Ultrasound to demonstrate complexity of effusion, and to guide thoracostomy
- C. CT to better characterize empyema, and to guide thoracostomy tube placement
- D. CT to better characterize opportunistic infection
- E. Upright radiograph to show fluid level in lung abscess



Correct Answer: B

Rationale

- A. Option A is not correct. Decubitus radiographs have limited value in large effusions, when the underlying lung is markedly consolidated.
- B. Option C is not correct. CT is less helpful than ultrasound to determine complexity of pleural collections, and is less cost-effective to guide treatment decisions [1-3].
- C. Option D is not correct. CT is mainly indicated in immunocompromized patients [4,5]. This patient had a normal immune system
- D. Option E is not correct. The radiograph was already taken upright, showing a pleural collection (empyema), not an intraparenchymal mass (abscess).

References

1. Ramnath Pediatrics 1998
2. Tan Kendrick Pediatr Radiol 2002
3. Kearney Clin Radiol 2000
4. Winer-Muram HT Radiology 1997
5. Barloon Chest 1991