24. An infant with recurrent pneumonia underwent a frontal chest radiograph (Fig 24-A) followed by contrast-enhanced CT (Figs 24-B, 24-C). Based on the patient's imaging findings, what is the most likely diagnosis?

A. Pulmonary sequestration
B. Congenital pulmonary airway malformation (CPAM)
C. Congenital lobar hyperinflation
D. Pulmonary arteriovenous malformation (AVM)

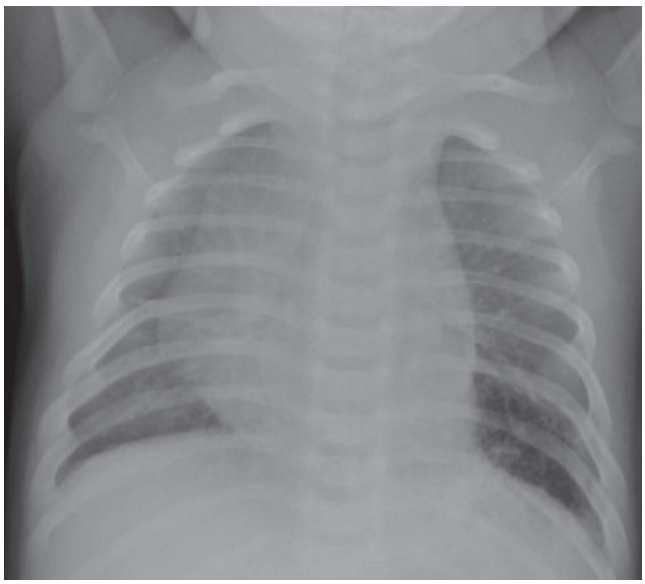

Figure 24-A. Frontal chest radiograph.

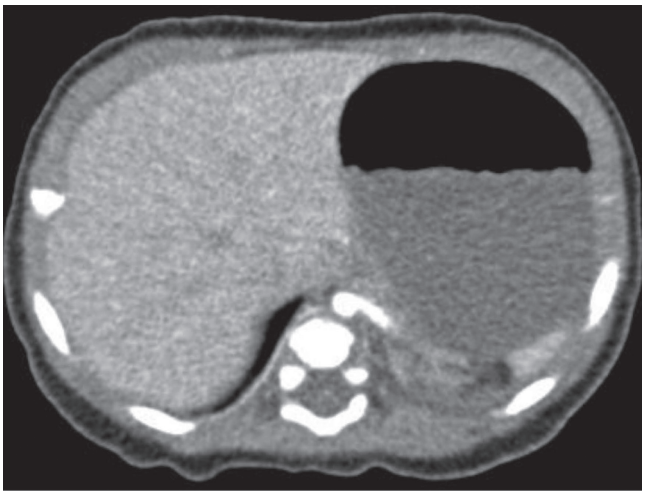

Figure 24-B. Contrast-enhanced axial CT image.

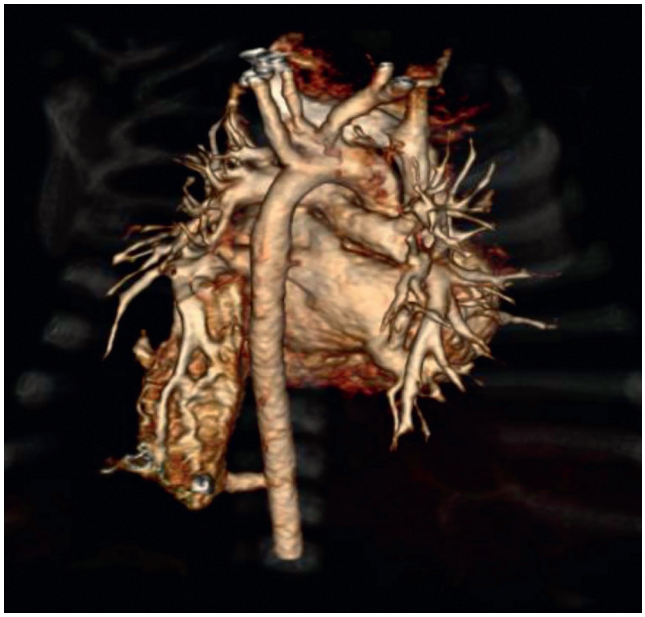

Figure 24-C. Posterior view of three-dimensional, volume-rendered image better shows a pulmonary sequestration with an associated anomalous artery arising from the descending aorta and venous drainage into the inferior pulmonary vein.

24. A. Pulmonary sequestration

Congenital lung malformations are a heterogeneous group of developmental anomalies affecting the lung parenchyma as well as arterial supply and venous drainage of the lung. In both asymptomatic and symptomatic pediatric patients with underlying congenital lung malformation, imaging assessment, particularly for surgical lesions, plays an important role. The current initial imaging modality of choice for evaluating congenital lung malformation is chest radiography. On chest radiographs, congenital lung malformation may present as a focal lung lesion or lucency or combination of both. Underlying associated vascular components of congenital lung malformation can be evaluated with cross-sectional imaging studies such as CT or MRI. Although MRI has an advantage over CT particularly in pediatric patients, due to lack of associated potentially harmful ionizing radiation, contrast-enhanced CT technique is the current cross-sectional imaging modality of choice after chest radiographs, due to its ability to provide essential anatomic information of both lung parenchymal and vascular anomalies associated with congenital lung malformation.

The frontal chest radiograph of this patient shows focal lung opacity (Fig 24-A) located in the retrocardiac region of the medial left lower lobe. The subsequently obtained contrast-enhanced axial CT image (Fig 24-B) demonstrates an anomalous artery arising from the descending aorta and delivering the blood supply to the left lower lobe lung lesion. The posterior view of the three-dimensional, volume-rendered CT image shows an anomalous artery arising from the descending aorta and venous drainage into the left inferior pulmonary vein, confirming the diagnosis of pulmonary sequestration (Fig 24-C). Pulmonary sequestration is a congenital lung malformation consisting of a portion of nonfunctioning lung that is not communicating with the adjacent tracheobronchial tree and that receives systemic arterial supply (choice A is correct). Once a diagnosis is made, surgical resection is the current management of choice particularly in symptomatic pediatric patients.

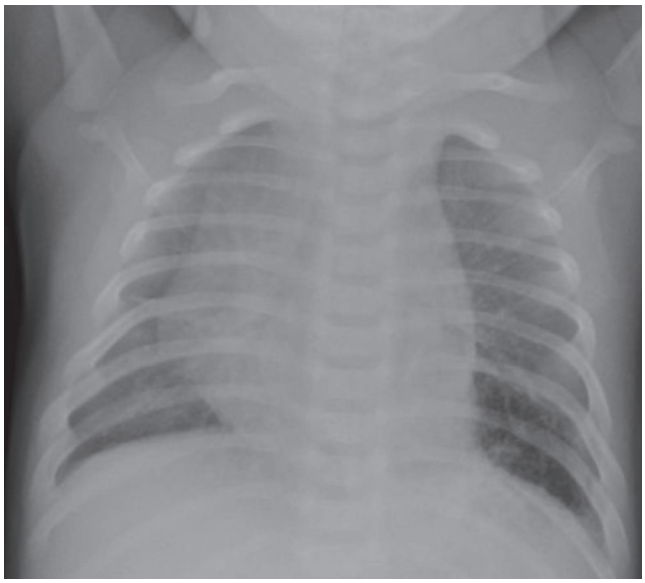

Figure 24-A. Frontal chest radiograph shows a focal opacity located in the medial left lower lobe, retrocardiac region.

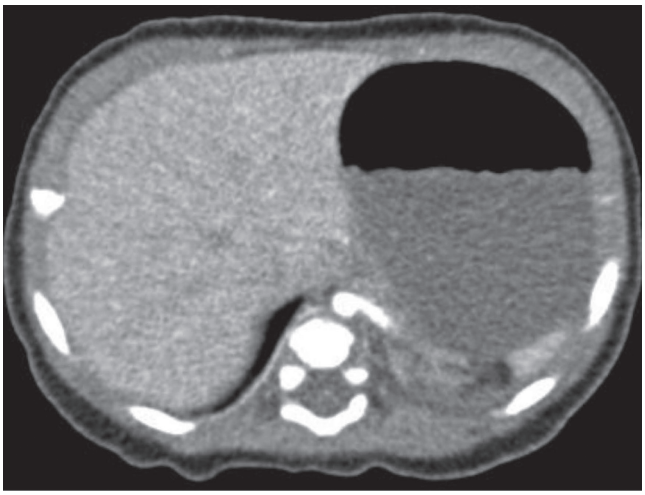

Figure 24-B. Contrast-enhanced axial CT image demonstrates an anomalous artery arising from the descending aorta and supplying blood to the malformed lung tissue located in the medial left lower lobe.

Congenital pulmonary airway malformations (CPAMs), previously known as congenital cystic adenomatoid malformations, are congenitally malformed lung tissues pathologically characterized by extensive overgrowth of the primary bronchioles. On imaging studies, CPAMs can be either cystic, solid, or a combination of both. Unlike pulmonary sequestration, arterial blood supply of CPAMs is from the pulmonary artery (choice B is incorrect).

Congenital lobar hyperinflation, previously known as congenital lobar emphysema, is a congenital lung lesion characterized by the overinflation and distension of one or more pulmonary lobes. Although there is no definite information regarding underlying causes of developing congenital lobar hyperinflation, it has been postulated that congenital lobar hyperinflation occurs due to either intrinsic or extrinsic bronchial wall narrowing with subsequent, distal air trapping. On imaging studies, a hyperinflated lobe with attenuated pulmonary vessels is typically seen. When it becomes large, it may also result in a mediastinal shift to the contralateral side. Unlike pulmonary sequestration, there is no anomalous systemic arterial supply (choice C is incorrect).

Pulmonary arteriovenous malformation (AVM) is a congenital vascular malformation characterized by a direct communication between a pulmonary artery and vein without an intervening capillary network. In pediatric patients, most pulmonary AVMs are congenital and may be associated with hereditary hemorrhagic telangiectasia (HHT), also known as Rendu-Osler-Weber syndrome. Pediatric patients with HHT often clinically present with the triad of epistaxis, mucocutaneous, or visceral telangiectasia, and family history of pulmonary AVM. On imaging studies, pulmonary AVM usually presents as a well-circumscribed serpiginous or lobulated opacity. Unlike pulmonary sequestration, anomalous arterial supply is from the pulmonary artery, which is directly connected to pulmonary vein via the nidus (choice D is incorrect).

Lee EY, Dorkin H, Vargas SO. Congenital pulmonary malformations in pediatric patients: review and update on etiology, classification, and imaging findings. *Radiol Clin North Am*. 2011;49(5):921-948.
Lee EY, Boiselle PM, Cleveland RH. Multidetector CT evaluation of congenital lung anomalies. *Radiology*. 2008;247(3):632-648.
Yikilmaz A, Lee EY. CT imaging of mass-like nonvascular pulmonary lesions in children. *Pediatr Radiol*. 2007;37(12):1253-1263.

25. A term infant is delivered vaginally following an uncomplicated pregnancy. Meconium is present in the amniotic fluid, and small amounts are suctioned from below the infant's vocal cords. Initially the baby is mildly tachypneic but well oxygenated on room air. Several hours after delivery, the infant is noted to be hypoxemic with increased work of breathing. A chest radiograph shows mild hyperexpansion and a few areas of patchy infiltrate bilaterally. Physical examination demonstrates a soft murmur at the left upper sternal border, but is otherwise unremarkable. Despite use of supplemental oxygen and nasal CPAP, the infant becomes progressively distressed. Echocardiography reveals a patent ductus arteriosus (PDA) and foramen ovale, but no structural defect.

Given this clinical scenario, which pattern of transcutaneous oxygen saturations would be expected from sensors placed on the right hand and the left foot?

Choices	Spo_2 Right Hand, %	Spo_2 Left Foot, %
A	90	82
B	96	97
C	84	91
D	82	82

25. A. Right hand: Spo_2 of 90%; left foot: Spo_2 of 82%

The patient described in the vignette has clinical signs of pulmonary arterial hypertension, most likely associated with meconium aspiration. In neonates who still have a PDA, this condition causes right-to-left shunting through the ductus and decreased oxygenation in the aortic blood flow below the level of the ductus. The oxygen saturation reading obtained from the right hand is considered to be "preductal," because the blood supply to the right upper extremity is derived from the right subclavian artery, which is fed from the aorta before the level of the ductus arteriosus. Blood delivered to the lower extremities has lower oxygen saturation, because it is mixed with blood shunted from the pulmonary arteries to the aorta through the open ductus. The difference in the preductal and postductal oxygen saturations can be used to measure the efficacy of therapy for pulmonary arterial hypertension in infants whose ductus arteriosus is known to be patent (choice A is correct).

A 96% Spo_2 reading from the right hand and a 97% Spo_2 reading from the left foot shows a minimal increase in the postductal oxygen saturation as compared to the preductal oxygen saturation. This seemingly anomalous finding most likely represents minor inaccuracies in the reading obtained by the oximeter, and is within the range of expected variation between similar units in simultaneous use (choice B is incorrect).

An 84% Spo_2 reading from the right hand and a 91% Spo_2 reading from the left foot shows the pattern of reversed differential cyanosis of the newborn, wherein the oxygen saturation in the lower extremities exceeds the preductal oxygen saturation. This unusual finding has been described in infants with a combination of congenital cardiac anomalies that include either transposition of the great arteries (TGA), PDA, and elevated pulmonary vascular resistance; or TGA, PDA, and preductal aortic interruption/coarctation. It can also be seen in patients with supracardiac total anomalous pulmonary venous return. In each of these conditions, highly oxygenated blood essentially bypasses the pulmonary arterial circulation and passes through a PDA directly into the aorta (choice C is incorrect).

An 82% Spo_2 reading from the right hand and an 82% Spo_2 reading from the left foot shows significant desaturation in both the preductal and postductal circulations, implying primary pulmonary pathology. An infant only a few hours old would be expected to retain a PDA, particularly in the setting of pulmonary hypertension, so this is not a likely finding for the clinical vignette described. In an older infant with known pulmonary hypertension, a change from the pattern seen in example A to this pattern might indicate that the ductus is closing (choice D is incorrect).

Roberts JD, Polaner DM, Lang P, Zapol WM. Inhaled nitric oxide in persistent pulmonary hypertension of the newborn. *Lancet*. 1992;340(8823):818-819.

Jegatheesan P, Song D, Angell C, Devarajan K, Govindaswami B. Oxygen saturation nomogram in newborns screened for critical congenital heart disease. *Pediatrics*. 2013;131(6):e1803-e1810.

Narang A, Bhakoo ON, Nair PMC, Bhandari V. Persistent pulmonary arterial hypertension of the newborn. *Indian J Pediatr*. 1992;59(6): 735-739.

Yap SH, Anania N, Alboliras ET, Lilien LD. Reversed differential cyanosis in the newborn: a clinical finding in the supracardiac total anomalous pulmonary venous connection. *Pediatr Cardiol*. 2009;30(3):359-362.