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Pulmonary Imaging Abnormalities in an Adult Case of Congenital Lobar Emphysema

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ABSTRACT

Congenital lobar emphysema is mainly diagnosed in infants, although rare cases are reported in adults. A 20-yr-old female with acute dyspnea, chest pain and left upper lobe (LUL) chest x-ray hyperlucency underwent 3He magnetic resonance imaging (MRI) for ventilation and apparent diffusion coefficient (ADC) measurements, as well as CT for emphysema and airway wall measurements. Forced expiratory volume in 1s, residual volume, and airways-resistance were abnormal, but there was normal carbon-monoxide-diffusing-capacity. The LUL relative area of the density histogram <-950 HU and airway morphology were highly abnormal compared with the other lobes and coincident with highly abnormal MRI-derived acinar duct dimensions. CT also identified bronchial atresia and congenital lobar emphysema as the source of symptoms in this case where there was also functional imaging evidence of collateral ventilation from the fissure (and not the abnormally terminated airway) into the emphysematous LUL.

CASE REPORT

CASE REPORT

A twenty-year-old female with a one-year, five-cigarette/day smoking history and no previous chronic respiratory disease presented with severe chest pain and shortness of breath. After meeting with a respirologist, the patient underwent a chest x-ray which showed localized left upper lobe (LUL) hyperlucency. Subsequent to this, in a follow-up visit to book pulmonary function tests, she was also referred to our pulmonary imaging research centre because of suspected CLE. Because of the unique pulmonary functional and micro-structural MRI measurements that can be made available using 3He MRI, it was thought that this novel information would help determine the etiology of symptoms, upon which a patient management and treatment plan could be initiated. After providing written informed consent to a Health Canada and local ethics board approved research protocol, the subject underwent two visits, 75 days apart that included spirometry, plethysmography and hyperpolarized 3He magnetic resonance imaging (MRI) to evaluate regional pulmonary ventilation [1] as well as parenchymal morphology using the 3He apparent diffusion coefficient (ADC) - a surrogate measurement of emphysema [2]. MRI was performed in breath-hold after inhalation of a 1L 3He/N2 gas mixture from functional residual capacity (FRC) using a fast-gradient-recalled-echo (FGRE) sequence, previously described

[3]. 3He MRI ventilation images were quantified to generate ventilation defect percent (VDP) [3] which is the volume of the lung not participating in ventilation, normalized to total lung volume. For the generation of 3He ADC, diffusion-weighted images were acquired as previously described [3]. To evaluate parenchymal microstructure, we adapted a method [2] whereby D_L and D_T were defined as the longitudinal and transverse diffusion coefficients in the acinar ducts respectively with R representing the outer radius and h the depth of the intra-acinar duct.

Thoracic CT was acquired once within twenty minutes of MRI using a 64-slice Lightspeed VCT scanner (GEHC, Milwaukee, WI, USA) as previously described [3]. Pulmonary Workstation 2.0 (VIDA Diagnostics, Iowa City, IA, USA) was used to generate emphysema measurements including the relative area of the lung with attenuation values <-950HU (RA950), low attenuation clusters (LAC), and as well airway measurements including airway wall area percent (WA%) and lumen area (LA).

Spirometry, plethysmography and diffusing capacity of carbon monoxide (DLCO) measurements showed there was modestly abnormal forced expiratory volume in one second (FEV1, V1:V2 70%:75%), and highly abnormal residual volume (RV, V1:V2 125%:150%), and airways resistance (Raw, V1:V2 185%:180%), but normal DLCO (V1:V2 100%:115%). Figure 2 shows CT low attenuating area (LAA) and low attenuation clusters (LAC) and 3He MRI ventilation imaging with a large LUL ventilation defect spatially coincident with the CT- and 3He MRI-defined emphysematous region. 3He MRI morphometric maps (higher intensity representing greater acinar duct dimensions) are also shown for the intra-acinar duct outer radius (R), depth (h), and inner radius (R-h). Quantitative LUL measurements showed abnormal RA950 (LUL: 13%, other lobes: 0%), LAC (LUL: -1.5, other lobes: -2.3 to -1.8) and VDP (LUL: 29%, other lobes: 1%-7%). For the LUL, acinar outer and inner radii were markedly abnormal (490µm and 310µm respectively) compared to the other lung lobes (300-310µm and 130-140µm respectively). CT airway measurements for the subsegmental bronchus leading to the LUL were abnormal (WA%: 50%, LA: 19mm2) compared to other airways of the same generation (WA%: 57%-66%, LA: 10mm2- 16mm2). In Figure 3, the CT-derived airway tree is shown in yellow co-registered with the 3He MRI ventilation image with the hyperlucent region outlined in white and the fissure between the LUL and LLL shown in black. The left upper subsegmental bronchus is abnormally terminated with the proximal end apparently disconnected from the rest of the airway tree. Relative dimensions for this abnormal airway and a similar generation normal airway are shown in schematic along with acinar duct morphological measurements for normal and abnormal LUL parenchyma estimated using 3He MRI and the Weibel model [4] assumptions.

DISCUSSION

Etiology and demographics

Congenital lobar emphysema (CLE) is a rare pulmonary abnormality that is typically detected in the neonatal period, although rare cases have been reported in early adulthood [5]. CLE affects males more than females (ratio of 3:1) and is commonly associated with bronchial abnormalities/obstruction and vascular dysfunction, resulting in ventilation-perfusion mismatch, hyperinflation and dyspnea [6]. Diagnosis is often based on x-ray computed tomography (CT) findings of hyperlucent and/or hyperinflated lung lobes along with malformations of the pulmonary vasculature or mediastinum. Here, we used imaging to quantitatively evaluate the function and morphology of alveolar and airway structures in a young adult with acute dyspnea and chest pain.

Clinical and imaging findings

Thoracic CT showed that there was a highly localized hyperlucent region in the LUL with a mediastinal shift to the contralateral right side likely related to LUL over-inflation - a common finding in CLE. Interestingly, we also observed focal obliteration of the proximal aspect of the tertiary bronchi serving the LUL apico-posterior segment (the LUL-LB1+2 bronchus). This leads to a lack of connection between this segment and the central airways although there was no evidence of localized endobronchial mucus plugging, foreign body or other masses resulting in extra-luminal compression. The distal segment of the apico-posterior bronchus was present but highly abnormal with a thinner wall (decreased WA%) and widening of the lumen (high LA) compared to similar airways in other regions, consistent with an occult atretic bronchus. The precise relationship between bronchial atresia and CLE is unknown although the timing and level of embryofetal airway obstruction may explain the variety of congenital lung lesions.

We also observed a markedly elevated (worse) LUL VDP compared to the other lobes and a heterogeneous pattern of 3He signal filling inwards from the edges of the emphysematous region of the apico-posterior region. We propose collateral ventilation into the emphysematous area as a potential mechanism for this finding, first described in excised emphysematous lungs [7] and COPD patients [8]. In emphysema, there is lower resistance to collateral flow that may involve the 1-2 micrometer pores of Kohn, the 30 micrometer channels described by Lambert, or the 80-150 micrometer interbronchiolar paths described by Martin [9].

In contrast to healthy lung tissue in the unaffected lobes, there was an elevated outer and inner radius of the LUL acinar ducts as previously shown in patients with emphysema. There was also an elevated acinar duct sleeve depth and this is a novel finding that contrasts with results in emphysema ex-smokers where decreased alveolar duct sleeve depths were reported.

Finally, although we observed significant airway and parenchymal abnormalities, airflow limitation was mild and DLCO was normal in this adult patient. There was evidence of gas trapping and very high airways resistance, both of which may be related to the atretic bronchus and slower time

constants for LUL emptying which may have been worsened by collateral ventilation. The increased airways resistance may also be driving collateral ventilation into the emphysematous lobe and hyperinflation- a potential source of severe symptoms in this patient. Other pathologies that should be considered when encountering hyperlucencies on x-ray or CT that resemble CLE (in infants or adults) include; bronchial atresia, congenital pulmonary airway malformations (CPAMs), bullous emphysema and spontaneous or traumatic pneumothorax.

Differential Diagnoses

Bronchial atresia is generally asymptomatic and most commonly diagnosed incidentally using x-ray or CT, similar to CLE. CLE often coexists with bronchial atresia making it difficult to differentiate these pathologies independently. In adult cases, both bronchial atresia and CLE show locally hyperinflated/emphysematous regions however, bronchial atresia patients exhibit mucoceles (club-like or rounded shape pathologies at the proximal ends of segmental or subsegmental bronchi) which are easily visualized on thoracic CT. Further investigation with bronchoscopy after the location of mucoceles may then identify blind-ending bronchi characteristic of bronchial atresia.

Congenital pulmonary airway malformations (CPAMs, also known as congenital cystic adenomatoid malformations (CCAMs)) appear on CT as cysts attached to segmental bronchi and are frequently observed concomitant with other congenital lung lesions such as bronchopulmonary sequestration or bronchial atresia. The diagnoses of CPAM typically occurs in the neonatal period and is sub-categorized into one of five subtypes with "type 3 CPAM" being the most similar to CLE because it affects at least one entire lobe. In prenatal cases, ultrasound most often identifies the presence of CPAM as hyperechoic lesions with microcystic patterns. If these malformations are not detected until adulthood, recurrent pulmonary infections are the most common finding that subsequently may result in incidental chest x-ray or CT findings.

Swyer-James syndrome, a pulmonary abnormality acquired after infection, may also resemble CLE on a chest x-ray and/or CT. Swyer-James, like CLE, appears as hyperlucency localized to one (or several) lobe(s) of the lung with the entire affected lung having markedly reduced parenchyma and vascular density. The key distinguishing factors that differentiate Swyer-James and CLE is an acquired form of segmental emphysema that arises as a sequelae of bronchopulmonary infections in childhood resulting in bronchiolitis obliterans and distal airspace destruction. Bronchiectasis on the affected side is a common finding in Swyer-James syndrome, while congenital heart disease is not typically associated with this condition [10]. Finally, CLE may resemble primary spontaneous pneumothorax (PSP) as usually occurs in young thin males without evidence of prior lung disease. Both CLE and PSP share a common feature in a prominent mediastinal shift towards the unaffected lung (opposite the hyerpucent lobe/region). The presence or absence of bronchovascular markings in the hyperlucent area is the main distinguishing factor between the two conditions on plain chest x-ray. Treatment approach is also different in the two conditions whereby severely symptomatic CLE requires surgical intervention with a lobectomy while large symptomatic pneumothraces require aspiration or chest tube drainage, which may worsen the outcome of CLE patients.

It is important to note that with the increasing use of chest CT, all these pulmonary pathologies may be observed in young adults and frequently, the exact genesis of the malformations may never be determined. This case underscores the clinical value of the regional structural and functional information that can be extracted from thoracic CT and 3He MRI. Merging functional information from 3He MRI with well-established structural measurements from CT allows for direct pulmonary structure-function assessments which can be used, as was shown here, to more fully characterize pulmonary abnormalities that result in severe symptoms.

Treatment and prognosis

Following the imaging investigation undertaken here the patient was counseled to stop smoking tobacco and marijuana and was booked for bi-annual visits with a respirologist that would include plethysmography measurements of gas trapping. In the case of worsening symptoms, the potential for lung volume resection will be discussed with this young female patient.

TEACHING POINT

In a 20 year old with acute severe chest pain and dyspnea, there was 3He MRI and CT evidence of congenital lobar emphysema in the LUL that was not evident using clinical spirometry or diffusing capacity breathing tests. In young adults with recent and sudden onset of chest pain and dyspnea, the presence and diagnosis of congenital lobar emphysema with or without bronchial atresia can be made using anatomical high resolution CT and functional/ventilation imaging using CT or MRI.

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FIGURES

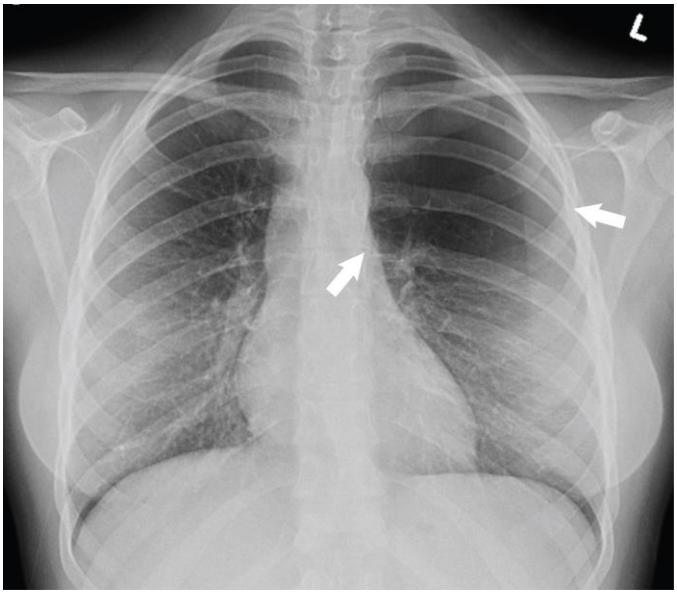
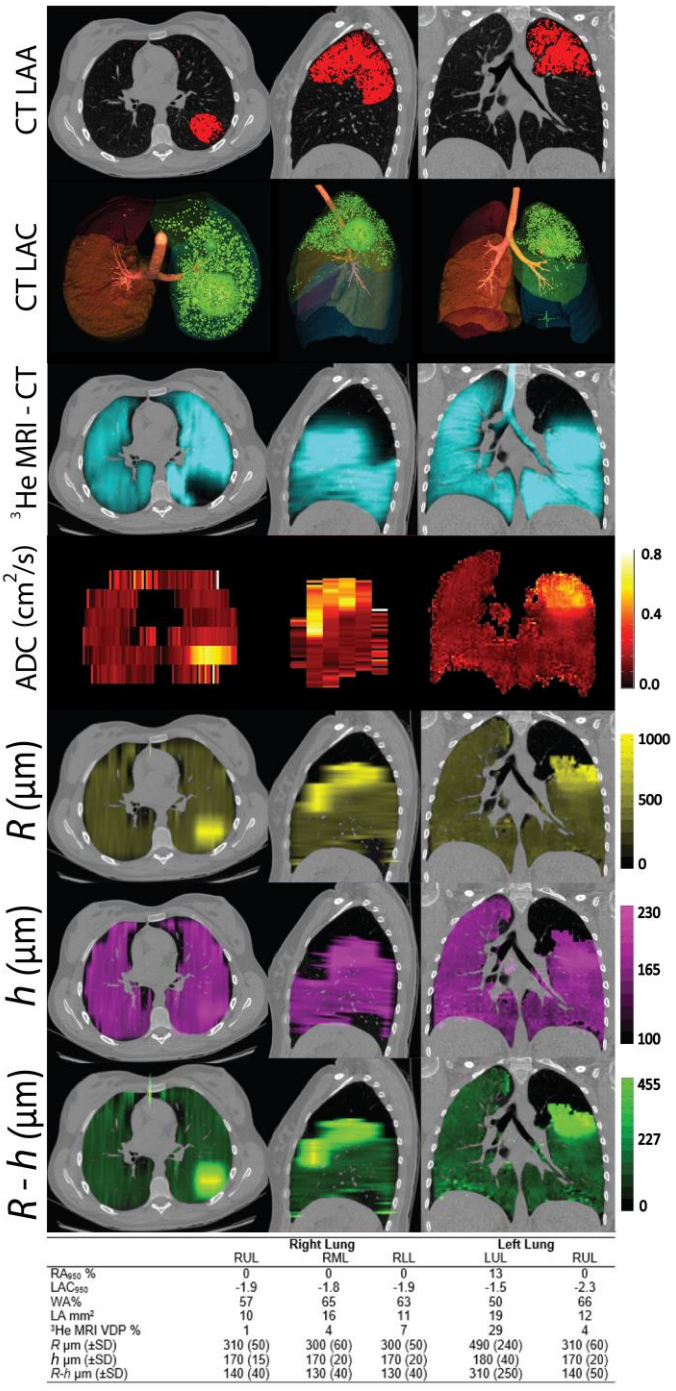


Figure 1: Anterior-poster chest X-ray of the 20-year old female patient with congenital lobar emphysema presented in this case. The emphysematous left upper lobe (LUL) is indicated with white arrows. The LUL has markedly reduced lung density and hyperinflation which has caused the mediastinum to shift onto the contralateral right lung.

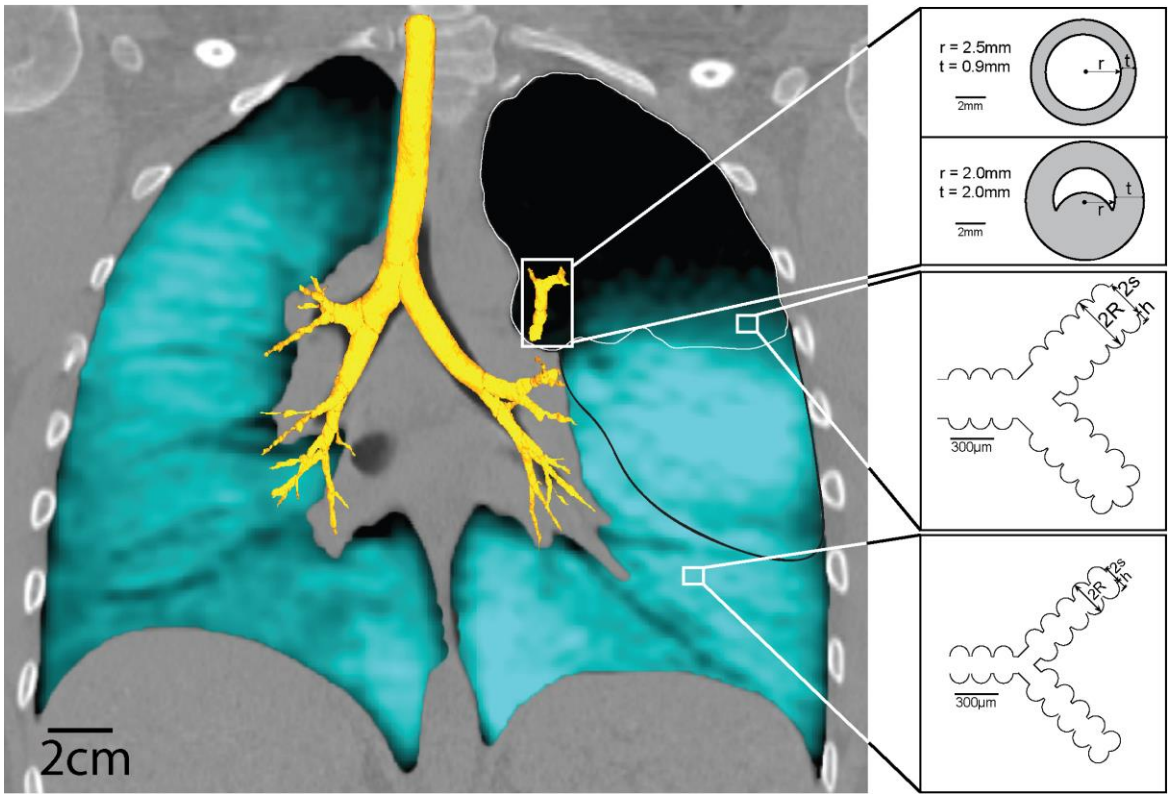


	Right Lung			Left Lung	
	RUL	RML	RLL	LUL	RUL
RA_{950} %	0	0	0	13	0
LAC_{950}	-1.9	-1.8	-1.9	-1.5	-2.3
WA%	57	65	63	50	66
LA mm²	10	16	11	19	12
^{3}He MRI VDP %	1	4	7	29	4
R µm (±SD)	310 (50)	300 (60)	300 (50)	490 (240)	310 (60)
h µm (±SD)	170 (15)	170 (20)	170 (20)	180 (40)	170 (20)
R-h µm (±SD)	140 (40)	130 (40)	130 (40)	310 (250)	140 (50)

Figure 2: Thoracic CT and 3He MRI measurements, co-registration and analysis in the axial, sagittal and coronal planes for the 20-year old female with congenital lobar emphysema presented in this case.
CT LAA: CT low attenuation area (LAA, in red, attenuation <-950HU) with the thoracic CT as the background (in grey-scale) shows a localized region of low density in the LUL, CT LAC: 3D low attenuation clusters (LAC) analysis generated using VIDA Pulmonary Workstation 2.0 shows the low density region observed on CT is predominated by a large bulla, 3He MRI-CT: 3He MRI static ventilation image (in blue) registered to thoracic CT (in grey-scale) show the spatial relationship between 3He ventilation and the emphysematous region in the LUL, 3He MRI ADC: 3He MRI apparent diffusion coefficient (ADC) maps for b=1.6cm2/s, where brighter yellow-green represents higher ADC values. The

ADCs are elevated (>0.40 cm2/s) in the LUL with other lung regions having normal ADC (0.15 - 0.25 cm2/s). R: intra-acinar duct radius generated from 3He MRI co-registered with thoracic CT (in grey-scale), h: intra-acinar duct depth generated from 3He MRI co-registered with thoracic CT (in grey-scale), R-h: intra-acinar duct inner radius co-registered with thoracic CT (in grey-scale). R, h and R-h maps show that dilation of the inner-acinar ducts is localized to the emphysematous region in the LUL.

Figure 3 (bottom): Registered CT airway tree with 3He MRI and CT for the 20 year-old female with congenital lobar emphysema presented in this case. The white contour outlines the emphysematous region in the left upper lobe (LUL), the black contour outlines the entire LUL. There is a noticeable mediastinal shift onto the contralateral right lung due to hyperinflation in the LUL. The subsegmental bronchus feeding into the LUL was abnormally terminated and not connected to the rest of the airway tree and dimensions of the resulting airway segment were abnormal (top panel). Acinar duct schematics for the abnormal LUL parenchyma (middle panel) and normal parenchyma (bottom panel) are shown to scale and were estimated using Weibel model assumptions.



Etiology	• Characterized by compression of lung tissue – emphysema, that is usually localized to one but can be several lung lobes • Usually the upper lobes are affected (>50% of cases) • Trapped gas within the localized emphysematous region causes displacement of the mediastinum into the opposite lung
Incidence	• Prevalence of 1 in 20,000 to 1 in 30,0000
Gender Ratio	• Affects males more than females 3:1
Age Predilection	• Nearly all cases present within the first year after birth but rare cases have been diagnosed in adulthood
Risk Factors	• Prenatal vascular abnormalities • Congenital heart disease (ventricular septal defects, ductus arteriosus)
Treatment	• Lobectomy is the accepted treatment for CLE • In older patients conservative management may be effective • In infants and neonates an emergency thoracotomy is sometimes performed
Prognosis	• Following surgical intervention, patients generally develop normal lung function without respiratory impairment in follow-up (>1 year)
Findings on Imaging	• Localized hyperlucency or regions of reduced density (<-950HU) on x-ray computed tomography (CT) scans • Mediastinal shift towards the opposite lung from the affected lobe • Tracheo-bronchial and abnormalities • Localized abnormalities to the pulmonary vasculature in the affected lung lobe

Table 1: Summary Table for Congenital Lobar Emphysema (CLE) and Tracheo-bronchial abnormalities

Disease	Clinical	Chest X-ray	CT	Other modalities
Bronchopulmonary causes				
Congenital Lobar Emphysema	• Dyspnea, tachypnea, wheezing, tachycardia and cyanosis are common symptoms. • Fifty percent symptomatic in the neonatal period.	• Hyperlucency of involved lobe, most commonly left upper lobe (LUL). • Herniation of hyperinflated lobe to opposite side with mediastinal shift and adjacent lobe collapse. • Flattening of ipsilateral diaphragm. • Scant lung markings are usually present within the radiolucent area.	• Localized hyperlucency and loss of parenchymal density in an individual lung lobe. • Attenuated sparse vasculature of the emphysematous lobe. Loss of distal airways in localized emphysematous lobe, compressive atelectasis of other lobes. • Abnormally narrowed bronchus or bronchial atresia may be present, however many cases do not have any demonstrable airway anomaly.	*MRI:* Prominent region of signal void on ^{3}He MRI ventilation images. Elevated apparent diffusion coefficients (ADC) on diffusion-weighted ^{3}He MR images. Abnormal outer and inner radii and acinar duct sleeve depth in localized emphysematous lobe
Bronchial Atresia	• Usually asymptomatic and diagnosed incidentally on x-ray in an older child or adult. • Since no communication exists with the normal tracheobronchial tree, infection is rare.	• Hilar-mass like shadows with hyperexpansion, oligemia and hyperlucency of the peripheral lung fields in the affected lobe.	• Presence of a mucocele (club-like or rounded in shape) with focal interruption of a lobar, segmental, or subsegmental bronchus. • Hyperinflation of the affected lobe or segment and emphysematous changes of the peripheral lung fields and occlusion of the bronchus central to the mucocele are diagnostic criteria.	*Bronchoscopy:* A blind-ending bronchus may be found in up to 50% of cases, which is pathognomonic. Helpful to exclude acquired causes of bronchial obstruction such as foreign body, strictures and tumors.
Congenital pulmonary airway malformations (CPAM)	• Rare cystic lung lesions arising from proliferation of tubular bronchial structures. • 5 subtypes with 80% diagnosed in the neonatal period. • In adulthood, recurrent pulmonary infection is the most common symptom.	• Localized patchy density representing the cystic mass with an area of hyperinflation and parenchymal oligemia. • Type 3 CPAM can involve an entire lobe or several lobes. • CPAMs that present during adulthood most commonly present in the lower lobes.	• Over-inflation with malformed multiple air cysts (irregular and thin walled) connected to the segmental bronchus and well demarcated from the normal lung parenchyma.	*Ultrasound:* Usually for pre-natal diagnosis revealing a homogenously hyperechoic lesion with microcystic pattern.
Swyer-James Syndrome (SJS)	• Commonly presents with a productive cough, wheezing, exertional dyspnea and recurrent pulmonary infections. • Other presenting features such as hemoptysis and cardiovascular anomalies are rare. • SJS is an acquired condition of segmental emphysema from bronchiolitis obliterans most commonly due to childhood pulmonary infections.	• Classically viewed as a unilateral process on chest x-ray although there are reports of bilateral disease. • Small or normal sized, hyperlucent lung or lobe with decreased bronchovascular markings, air trapping on expiratory radiograph, small ipsilateral hilar shadow and pulmonary vessels are the main features. • Hyperinflation and mediastinal shift towards the contralateral lung may be present. • Evidence of bronchiectasis and scarring may also be noted.	• Lobar hyperlucency and loss of attenuation of lung parenchyma on the affected side, usually smaller in size with sparse bronchovascular markings and small ipsilateral pulmonary artery is characteristic. • Decreased number of bronchial subdivisions and pulmonary artery branches as well as features of bronchiectasis may be present. • CT more frequently demonstrates multi-lobar or bilateral lung involvement, however, this is usually asymmetric with one lobe or lung being more hyperlucent.	*Magnetic Resonance Angiography (MRA):* Smaller pulmonary artery and its diminished branches peripherally on the affected radiolucent side with a 'pruned tree appearance' is apparent. On its own, this modality is not diagnostic but has the advantage of being fast, sensitive with no radiation exposure that helps support the diagnosis. Radionuclide lung scan: Demonstrates matched ventilation-perfusion mismatch of the affected segments (which helps differentiate from other causes of a unilateral hyperlucent lung with a vascular cause or bronchial obstruction).
Others: Airways obstruction due to bronchial compression from extrinsic factors (cardiomegaly, lymphadenopathy), endobronchial obstruction (tumor, foreign body, mucus plug) and obliterative bronchiolitis, Unilateral bulla/bullae and post-pneumonectomy are other differentials.				

Table 2: Differential Diagnosis Table for Congenital Lobar Emphysema

Disease	Clinical	Chest X-ray	CT	Other modalities
Pleural space causes				
Pneumothorax	• Dyspnea, tachypnea, tachycardia, cyanosis, decreased breath sounds, tracheal deviation in tension pneumothorax. • May be spontaneous, traumatic or iatrogenic.	• Visceral pleural line (and lung) is separated from parietal pleural (and chest wall) with hyperlucency of gas in between. • Tracheal deviation and mediastinal shift maybe present.	• Most sensitive imaging modality to detect pneumothorax. • Can detect small pneumothoraces, hydropneumothorax, loculated pneumothorax, bronchopleural fistulas, underlying emphysema/bullous lung diseases.	*Ultrasound:* Used in point-of-care situations e.g.: emergency department/ICU. Absence of 'lung sliding', seen as a shimmering line due to sliding of visceral and parietal pleural relative to each other is suspicious for a pneumothorax.
Others: A contralateral pleural effusion may sometimes appear as a hyperlucent hemithorax				

Table 2: Differential Diagnosis Table for Congenital Lobar Emphysema (continued)

ABBREVIATIONS

3He MRI: Hyperpolarized helium-3 magnetic resonance imaging
ADC: Apparent diffusion coefficient
CLE: Congenital lobar emphysema
CPAM: Congenital pulmonary airway malformations
CT: Computed tomography
DL: Longitudinal diffusion coefficient of acinar ducts
DLCO: Diffusing capacity of the lung for carbon monoxide
DT: Transverse diffusion coefficient of the acinar ducts
FEV1: Forced expiratory volume in one second
FGRE: Fast gradient recalled echo sequence
FRC: Functional residual capacity
h: Depth of the intra-acinar ducts
LA: Airway lumen area
LAA: Low attenuation areas
LAC: Low attenuation clusters
LLL: Left lower lobe
LUL: Left upper lobe
LUL-LB1+1: Left upper lobe apico-posterior segment
R: Outer radius of the intra-acinar ducts
RA950: Relative area of the lung with attenuation values <-950HU
Raw: Airways resistance
VDP: Ventilation defect percent
WA: Airway wall area

KEYWORDS

Congenital Lobar Emphysema; Bronchial Atresia; Hyperpolarized 3He; Magnetic Resonance Imaging; Computed Tomography; Emphysema; Airways Disease

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