Diagnostic approach to
Cystic lung disease for radiologists

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- Incidental cysts
- Pneumatocele
- Congenital anomaly
- Congenital pulmonary airway malformation (CPAM)
- Bronchogenic cyst

**Diffuse cystic disease**

Without other radiologic abnormality

Cystic disease only

- Lymphangioleiomyomatosis (LAM)
- Birt-Hogg Dube syndrome (BHD)
- Metastatic cancer

Associated with nodules

- Pulmonary Langerhans cell histiocytosis (PLCH)
- Amyloidosis

Associated with GGO

- Lymphoid interstitial pneumonia (LIP)
- Desquamative Interstitial pneumonia (DIP)
- Pneumocystis Jirovecii pneumonia (PCP)
Introduction
**Definition**

**Cyst**
- Round air space defined by an epithelial or fibrous outer wall

**Radiology**
- Round parenchymal lucency or low attenuation area with a well-defined interface with normal lung – Usually thin wall (<2-3 mm), containing air, occasionally fluid or solid materials

**vs. Cavity**
- Gas-filled space with develops in an area of pulmonary consolidation, mass or nodule
- Usually relatively thick wall (>4 mm)

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Figure 1a. A small air-cyst in RUL

Figure 1b. Pneumonia with large cavity in RUL
Definition

Cystic lung disease

- **Spectral disease entity associated with** simple or multiple radiolucency
  area (cystic) surrounded by discrete wall, not associated with cavity

- **Clinical manifestation**
  - Usually present with no symptom
  - Nonspecific symptom: cough, shortness of breath.
  - Pneumothorax → shortness of breath

- **Diagnosis**
  - Clinical + Radiological + Pathological correlation!!

Cysts; location, number, distribution, associated CT features (GGO, nodules)
**Approach to cystic lung disease**

1. **Step 1: True cysts?**
   - No
     - Cavitary disease
     - Centrilobular emphysema
     - Bullae
     - Honeycombing
   - Yes

2. **Step 2: Localized or diffuse?**
   - Localized
     - Incidental cysts
     - Pneumatocele
     - Congenital anomaly - CPAM - Bronchogenic cyst
   - Diffuse
     - LAM
     - BHD
     - Malignancy

3. **Step 3: Ancillary feature?**
   - Yes
     - PLCH
     - Amyloid
   - No
     - LIP
     - DIP
     - PCP

4. **Step 4: What kind of feature?**
   - with nodules
     - PLCH
     - Amyloid
   - with GGO
     - LIP
     - DIP
     - PCP

5. **Step 5: Distribution & Shape**
Step 1. Are they true cysts?
Step 1: Are these true cysts?

No

Cavitary disease

Centrilobular emphysema

Bullae

Honeycombing
Cavitary disease

- **Cavity**: Gas-filled space, seen as a lucency or low-attenuation area
  - With a wall thickness of > 4 mm
  - Noted within pulmonary consolidation, a mass, or a nodule
- Usually produced by the expulsion or drainage of necrotic portion of the lesion via bronchial tree
- Example: Infection, cavitary metastasis

*Figure 1B. Pneumonia with large cavity in RUL*
Mimicking cystic disease

Cavitary disease

Figure 2. M/52 Patient with colon cancer. Serial follow up CT show cavitary metastatic nodule (arrow) in LLL

Figure 3. M/52 Patient with colon cancer. Serial follow up CT show cavitary metastatic nodule in LLL
Mimicking cystic disease

**Centrilobular emphysema**

- *Destroyed centrilobular alveolar walls* and enlargement of respiratory bronchioles and associated alveoli
- *Centrilobular lucencies that lacks discrete walls*
- Predilection for *upper lung zones* / *Presence of central core vessel*
- Clinical information: smoker, COPD, PFT-obstruction

Mimicking thin walled cystic lesion - Difficult to distinguish

**Figure 4. M/52 Dyspnea**

Usually centrilobular emphysema lack discrete wall
Mimicking cystic disease

Centrilobular emphysema
– can mimic cystic lung disease

Figure 5. F/70 Dyspnea, hemoptysis (onset: 1 month)
(a-b) Sometimes, centrilobular emphysema may mimic thin-walled cystic lesion
(c-d) VATS biopsy was done. Pathologically confirmed destructed alveolar wall, suggesting centrilobular emphysema
Mimicking cystic disease

Bulla(e)

- Airspace measuring more than 1cm, sharply demarcated by a thin wall

- Radiology: rounded focal lucency or area of decreased attenuation more than 1cm, bounded by a thin, almost imperceptible wall
  - Usually accompanied by emphysematous change

- Clinical; Asymptomatic (m/c) – enlarged over time and compress normal lung resulting in restrictive physiology / spontaneous pneumothorax
  - Associated with Ehlers-Danlos, Marfan syndrome...
Mimicking cystic disease

Figure 6. M/69
(a) Focal lucent area bounded by a thin wall, suggestive of bulla in RLL.
(b) Follow up CT shows increased size of bulla in RLL
Mimicking cystic disease

Honeycombing

- Results from advanced and irreversible fibrotic lung disease

- Cysts that typically **3-10mm in diameter, clustered** (the walls of the cyst abut each other), with thick walls (1-3mm or larger)

  - With **architectural distortion and reticulation of intervening lung**

  - **Subpleural and well-defined walls**
Figure 7. M/68 Dyspnea
(a) Chest radiography show reticular opacities in both lower lung, periphery.
(b-c) Chest CT show reticular density with honeycombing, suggestive of UIP pattern.
(d-e) VATS lung biopsy shows UIP pattern with honeycomb change.
Step 2. Localized or diffuse?

Localized cystic lung disease
Localized cystic lung disease

Cystic lung disease

Step 1: Are these true cysts?

Yes

Step 2: Localized or diffuse?

Localized

- Incidental cyst
- Pneumatoceles
- Congenital anomaly
- CPAM
- Bronchogenic cyst
Localized cystic lung disease

Incidental air-cyst

- 25% of patient aged > 75 years (normal aging process)
- Clinical: Never caused symptoms

Smokers=non-smokers, distributed uniformly in all lobes
- Also be the remnant of a previous infection or trauma

- Absence of suspicious clinical feature → further evaluation unnecessary

Figure 8. M/71 Stomach cancer patient
Chest CT show incidental cyst (well-defined radiolucent area with thin wall)
Localized cystic lung disease

Pneumatocele

- **Transient, thin-walled, gas-filled space in the lung**
- **Caused by acute pneumonia, trauma, aspiration of hydrocarbon**
- **Mechanism**: parenchymal necrosis & check-valve airway obstruction
- **Vs. cyst** → transient nature, history such as pneumonia or trauma

**Figure 9.** M/22 Traffic accident
(a-b) Initial chest CT show focal thin-walled cystic space with adjacent GGO; pneumatocele and lung contusion
(c) Follow up chest CT after 1month disappeared cystic lesion which is characteristic of pneumatocele
Localized cystic disease

Congenital pulmonary airway malformation (CPAM)

- **Multicystic mass of pulmonary tissue** with an abnormal proliferation of bronchial structures

- **Pathology**: Overgrowth of bronchioles
  
  Almost complete suppression of alveolar development 7th-10th wk

  - Type I: Variable-size cysts, with at least one dominant cyst (>2 cm), most common form
  
  - Type II: Smaller, more uniform cysts <2 cm in diameter (10% to 15%)
  
  - Type III: Solid mass composed of bronchoalveolar microcysts (causes death at birth)

- Usually involves a single lobe and connected to the bronchus

- **Normal arterial supply and venous drainage**

- **Variable history and prognosis (depend on size rather than histologic type)**
Localized cystic disease

Congenital pulmonary airway malformation (CPAM)

- **Imaging findings**
  - Usually multiple air-filled thin-walled cysts that vary in size
    - Type III: Soft tissue density mass
  - Air-fluid level maybe seen with or without superimposed infection

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<th>Type II</th>
<th>Type III</th>
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<td>75%</td>
<td>10-15%</td>
<td>5%</td>
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<td>Large (&gt;2cm), air-filled, Multicystic lesion</td>
<td>Smaller (&lt;2cm), more uniform cysts / Associated with severe malformation (renal agenesis, dysgenesis)</td>
<td>Microscopic (&lt;3-5mm) cysts, present as a solid mass / Stillborn with polyhydroamnios or pulmonary hypoplasia</td>
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Localized cystic disease

Congenital pulmonary airway malformation (CPAM), type I

Figure 10. F/18 Abnormal chest imaging
(a) Chest radiography show cystic lesion in LLL.
(b-d) Chest CT show multicystic lesion in LLL (largest cyst size is larger than 2cm)
⇒ LLL lobectomy confirmed congenital cystic adenomatoid malformation, type I
Localized cystic disease

Congenital pulmonary airway malformation (CPAM), type II

Figure 11. M/23 months Incidental abnormal finding in antenatal ultrasound
(a) Chest radiography show hyperlucent lesion in RLL.
(b-e) Chest CT show multicystic lesion in RLL (largest cyst size is smaller than 2cm), suggestive of CPAM, type II
Localized cystic disease

Bronchogenic cyst

- Abnormal budding of the tracheobronchial tree during lung development
- Usually present as middle mediastinal masses
  - Also present as lower lobe intraparenchymal masses (1/3)
- Commonly filled with fluid, air-filled intraparenchymal cysts are rare
- CT: cyst with well-defined boundaries and water or soft tissue density

- Internal air-density
- Internal water or soft-tissue density
Localized cystic disease

Bronchogenic cyst

**Figure 12.** F/30 Abnormal chest imaging

(a) Chest radiography shows a 4.2cm sized smooth marginated ovoid soft tissue mass in RLL.
(b-d) Chest CT shows well-defined fluid-attenuated mass in RLL, suggestive of bronchogenic cyst
(e) Pathology confirmed bronchogenic cyst (H&E staining x100)
Localized cystic disease

Bronchogenic cyst

Figure 13. F/55 Mild dyspnea
(a) Chest radiography shows well-defined cystic lesion in RLL.
(b-c) Chest CT shows well-defined thin-walled air-density lesion in RLL, suggestive of bronchogenic cyst.
(d) VATS biopsy confirmed bronchogenic cyst.
Step 3. Ancillary features?

Diffuse cystic lung disease
- Without other radiologic abnormality
Diffuse cystic lung disease *without other radiologic abnormality*

**Cystic lung disease**

**Step 1:** Are these true cysts?  
- Yes

**Step 2:** Localized or diffuse?  
- Diffuse

**Step 3:** Ancillary features?  
- No

**Diffuse cystic lung disease**  
*without ancillary features*

- LAM
- BHD
- Cystic metastasis
**Diffuse cystic lung disease** without other radiologic abnormality

**Lymphangioleiomyomatosis (LAM)**

- **Rare interstitial lung disease by** proliferation of smooth muscle cells
- **Clinical history:** Dyspnea, cough, spontaneous pneumothorax...
  - Women, reproductive age (post menopausal: HRT)
  - Associated with tuberous sclerosis (TSC-LAM), sporadical
- **Extrapulmonary manifestation:** Renal angiomyolipoma, chylous ascites, uterine leiomyoma, abdomen & pelvis lymphangioleiomyoma

*Figure 14.*
(a) Grossly, multiple uniform cysts are scattered entire lung
(b) Microscopically, multiple cysts surrounded by bundles of smooth muscles (H&E staining x40)
**Diffuse cystic lung disease** without other radiologic abnormality

**Lymphangioleiomyomatosis (LAM)**

- **Imaging findings**
  - Diffuse parenchymal cysts; thin walled (up to <4mm), uniform in size
  - No zonal predominance (distributed throughout the lung)
  - Chylothorax (23%), Pneumothorax (69%)
  - TSC-LAM: diffuse nodular change along cysts

*Figure 15. F/28 Dyspnea – VATS biopsy confirmed LAM*
*Chest CT shows multiple uniform sized cysts distributed throughout the lungs (involving costophernic angle)*
Figure 16. F/35 Patient with tuberous sclerosis.

(a) Multiple thin walled uniform sized cysts with tiny nodules in both lungs, suggestive of LAM.

(b) Fat containing enhancing mass in both kidneys, suggestive of angiomyolipma.

(c) Multiple sclerotic lesions in bony thorax.
**Diffuse cystic lung disease** *without other radiologic abnormality*

**Multiple – Birt Hogg Dube syndrome (BHD)**

- **Autosomal-dominant** *(17p11.2) multiorgan systemic disorder*
- **Cutaneous lesions** *(fibrofolliculomas), renal tumors, pneumothorax*
- **Diagnosis**: One major criteria or two minor criteria!

### Major criteria

- At least fibrofolliculomas or trichodiscomas, at least one histologically confirmed, of adult onset
- Pathogenic FLCN germline mutation

### Minor criteria

- Multiple lung cysts: bilateral basally located lung cysts with no other apparent cause, with or without spontaneous primary pneumothorax
- Renal cancer: early onset (<50 years) or multifocal or bilateral renal cancer, or renal cancer of mixed chromophobe and oncocytic histology
- A first-degree relative with BHD
Diffuse cystic lung disease without other radiologic abnormality

Multiple – Birt Hogg Dube syndrome (BHD)

*Imaging findings*

- Well-defined cysts, vary in size and shape (>80%)
- Predominantly seen in peripheral lung zones at base, along mediastinum
- Either abut or encase the proximal portion of the lower pulmonary vein
- Spontaneous pneumothorax (24%)

*Figure 17.* F/48 Dyspnea (onset: 15 days ago)
(a) Patient with spontaneous pneumothorax in right.
(b-c) Chest CT shows variable size and shaped well-defined cysts, suggestive of BHD
**Diffuse cystic lung disease without other radiologic abnormality**

**Multiple – Birt Hogg Dube syndrome (BHD)**

**Figure 18.** F/32 Incidental finding (cancer work up)
(a) No abnormal finding in chest radiography.
(b-c) Chest CT shows variable size and shaped well-defined cysts along mediastinum, and lower lung
(d) Abdomen CT shows a heterogeneous well-enhancing mass in right kidney, suggestive of RCC
(e) Right partial nephrectomy confirmed clear cell type, RCC
### Diffuse cystic lung disease without other radiologic abnormality

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<table>
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<tr>
<th><strong>LAM</strong></th>
<th><strong>BHD</strong></th>
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<tr>
<td>Diffuse, many cysts</td>
<td>Lower part (+)</td>
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<tr>
<td>Small to medium</td>
<td>Large, multiseptated</td>
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<tr>
<td>More uniform</td>
<td>Various</td>
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* Involve the lungs and kidneys
**Diffuse cystic lung disease** without other radiologic abnormality

**Multiple – cystic metastasis**

- If cystic lung lesion in patient with known malignancy: D/D metastasis
  - Angiosarcoma, squamous cell cancer (esp. head and neck primary), Primary lung cancer (esp. adenocarcinoma)...

- **CT:** Multiple cysts - random distribution / peripheral, basal, subpleural

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**Figure 19-i.** M/58 Dyspnea, s/p Scalp wide excision for angiosarcoma  
(a-b) Chest CT shows multiple variable sized cystic lesion in both lungs, suggestive of cystic metastasis of angiosarcoma. And left pneumothorax is noted.
**Diffuse cystic lung disease** without other radiologic abnormality

Multiple – cystic metastasis

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*Figure 19-ii (continues). Treated with weekly paclitaxel.*

*Follow up chest CT shows decreased size and numbers of cystic metastasis.*
Step 4. What kind of feature?

Diffuse cystic lung disease
- With other radiologic abnormality
Diffuse cystic lung disease with other radiologic abnormality

Cystic lung disease

Step 1: Are these true cysts? Yes

Step 2: Localized or diffuse? Diffuse

Step 3: Ancillary features? Yes

Step 4: What kind of feature?

Associated with nodules:
- Pulmonary Langerhans cell histiocytosis (PLCH)
- Amyloidosis

Associated with GGO:
- Lymphoid interstitial pneumonia (LIP)
- Desquamative interstitial pneumonia (DIP)
- Pneumocystis jirovecii pneumonia (PCP)
Pulmonary Langerhans cell histiocytosis (PLCH)

- **Abnormal peribronchial proliferation of monoclonal Langerhans cells**

- **Clinical:** Strong association with cigarette smoking (>95%) - cytokine
  
  20-40 years old / M=F
  
  Sx: Asymptomatic (25%) - cough, dyspnea, pneumothorax (25%)
  
  Associated with malignant neoplasm (Hodgkin lymphoma)

- **Treatment:** Smoking cessation > corticosteroid > CTx or lung transplantation

- **Pathology**
  
  - Nodules of densely packed cells
  
  - Fibroblastic proliferation replacing nodules in a centripetal fashion
  
  - Central bronchial dilation
  
  - Coalescence of alveolar spaces
  
  - LCH cells absent
  
  - Only a fibrous, stellate scar remains

**Temporal heterogeneity**
Pulmonary Langerhans cell histiocytosis (PLCH)

**Imaging**

- **Early stage**
  - Small (1-10mm) irregular nodules
  - Bilateral, symmetric
  - Upper, middle lung zone
  - Sparing lung bases, CPA

- **Later stage**
  - Cystic degeneration
    - Round/oval ~ bizarre, thin walled
  - Associated with nodules
  - Upper lung zone

**Temporal heterogeneity**

- All cases, pathologically confirmed PLCH.

**Figure 20.**
- (a) M/33. Chest CT shows small nodules in BULs
- (b) M/29. Chest CT shows bizarre shape cysts with small nodules in both lungs, mainly upper lungs
- (c) M/35. Chest CT shows variable shape cysts in both lungs, mainly upper lungs

→ All cases, pathologically confirmed PLCH.
Diffuse cystic lung disease with nodules

Pulmonary Langerhans cell histiocytosis (PLCH)

Figure 21. M/22 Abnormal imaging finding from abdomen CT, current smoker (1.25ppd/9years)
(a-c) Chest CT shows multiple bizarre shape, thin-walled cysts with small irregular nodules in both lungs, sparing lung bases and CPA, suggestive of PLCH
(d-e) LUL wedge resection confirmed PLCH (f) Immunohistochemistry CD1a staining positive
**Amyloidosis**

- Abnormal deposition of **soluble plasma proteins** within the extracellular space in an **abnormal insoluble fibrillar form**

- **Systemic (80-90%) / Localized (10-20%)**
  - Primary (Associated MM) vs Secondary (Associated with RA, TB, Crohn’s disease)

- **Clinical;** Lung involvement – **6th decade, associated with Sjogren’s syndrome,** lymphoproliferative disease, MALT lymphoma

- **Type**
  - Tracheobronchial form: focal, diffuse tracheal narrowing
  - Nodular form: lung parenchymal nodules
  - Diffuse form: miliary nodules, reticulonodular opacities, honeycombing
**Amyloidosis**

- **Imaging**
  - Lung parenchymal involvement: **Nodules, interlobular septal thickening, GGO**
  - Cyst: rare, most often described with localized amyloidosis (Sjogren associated)

  Narrowing of airway due to inflammatory cells → check-valve mechanism

  Thin-walled, predominantly peripheral, associated with nodules


**Diffuse cystic lung disease with nodules**

**Amyloidosis**

Figure 22. F/66 Hemoptysis

(a–e) Chest radiography and CT shows variable sized nodules with internal calcification and thin-walled cysts

(f) Fluoroscopy guided needle biopsy confirmed amyloidosis
Lymphoid interstitial pneumonia (LIP)

- A rare benign polyclonal lymphoproliferative disorder of the lung parenchyma → More extensive & diffuse than follicular bronchiolitis
- Clinical; Associated with autoimmune disease, chronic infections, and several miscellaneous conditions, Dysproteinemia (60%)
  30-60 years old / F > M

- Pathology
  - Infiltration of polyclonal lymphocyte (predominantly T cells) into the alveolar interstitium

- Corticosteroid + Underlying disease control
- 1/3 – progressive (Death; infection...)
- Association to lymphoma
  - Transformation (controversial), combined
Diffuse cystic lung disease with GGO

Lymphoid interstitial pneumonia (LIP)

- Imaging
  - Ball-valve mechanism
  - Radiography: often normal; lower lung reticular and nodular opacities
  - CT: Perivascular cysts, centrilobular nodules, GGO - lower lung distribution
    - interlobular septal thickening, lymphadenopathy

Figure 23. F/51 Patient with Sjögren’s syndrome
GGO and centrilobular nodules in both lungs and several air-cyst in RML and RLL: possibility of LIP
Diffuse cystic lung disease with GGO

Lymphoid interstitial pneumonia (LIP)

Figure 24. F/63 Chronic cough for 5 months
Reticular density, GGO in BLL with several air-cysts, suggestive of LIP

 Courtesy to http://kstr.radiology.or.kr/weekly (case #729)
Desquamative interstitial pneumonia (DIP)

- **Idiopathic interstitial pneumonia – major – smoking related IIPs**
- **Clinical;** Strong association with cigarette smoking (90%) + others
  
  40-60 years old / M>F (2:1)
  
  Sx: nonspecific (shortness of breath, cough)

- **Pathology**
  
  - Increased number of pigmented macrophages evenly dispersed within the alveolar spaces
  
  - Alveolar septal thickening by diffuse fibrosis and mild interstitial inflammation

- **Treatment;** Smoking cessation

**Diffuse cystic lung disease with GGO**

Inhalation of inorganic particles, sirolimus treatment, RA
Desquamative interstitial pneumonia (DIP)

- **Imaging**
  - **Radiography:** Basilar reticulation & ill-defined airspace opacity
  - **CT:** GGO (80%), Reticular opacity (60%) – Basal & peripheral predominance
    - Small(<2cm), well-defined thin-walled lung cysts
      - Cysts within GGO!!

**Figure 25. M/52 Smoker**

Chest CT show ground glass opacity with tiny air-cysts in both lungs, mainly BLL, suggestive of DIP pattern. Lung wedge resection confirmed DIP.
Desquamative interstitial pneumonia (DIP)

Figure 26. M/69 Cough (2months) Smoker
Chest CT show ground glass opacity with internal tiny air-cysts in both lungs, suggestive of DIP pattern. Lung biopsy resection confirmed DIP.
Pneumocystis jirovecii pneumonia (PCP)

- **Fungal infection** (*pneumocystis jiroveci* – infects humans)
- **Clinical**: Strong association with immunocompromised patient – HIV/AIDS
  - 60-80% of AIDS patients at least one episode
  - CD4 lymphocyte count < 200 cells/μL
  - Symptom: Fever, shortness of breath, non-productive cough
  - Indolent course for HIV/AIDS, other immunosuppressive- acute course

- **Diagnosis**: PCR testing !!
  - Identification on stains of respiratory secretion

- **Treatment**: TMP-SMX

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**Figure 27. Pneumocystis jirovecii**
Gomori methenamine silver (GMS) stain shows grouped PCP
Pneumocystis jirovecii pneumonia (PCP)

- **Imaging**
  - **Ground-glass attenuation:** central, perihilar distribution / diffuse
  - **Interlobular septal thickening**
  - **Thin-walled cysts (30%)**: multiple, upper lobe predominance, variation in size, shape, wall thickness → increased pneumothorax

*Figure 28. M/51* Dyspnea (onset: 1 month ago). Patient diagnosed HIV positive 5 years ago
(a, b-c): Chest radiography and CT show diffuse GGO in both lungs, mainly central portion. PCR test confirmed PCP.
(d) Patient treated with TMP-SMX. After 1 month follow up radiography improved bilateral GGO.
Figure 29. M/36 Fever, cough, sputum, and dyspnea.
(a, b): Chest CT shows diffuse GGO in both lungs, peripheral sparing. Small thin-walled cysts are seen in BLLs. Patient with confirmed PCP.
Step 5. Distribution & shape?
Distribution & shape

**Distribution**

- **Upper dominancy**
- **Lower dominancy**
- **No zonal predilection**
- **Peripheral dominancy**

**<PLCH>**
- Upper, middle lung zone
- Sparing lung bases, PA
- Bizarre shape

**<PCP>**
- Upper lobe predominance, PA
- Variable size, shape, wall thickness
Distribution & shape

Distribution

Upper dominancy

Lower dominancy

No zonal predilection

Peripheral dominancy

Congenital anomaly: CPAM, Bronchogenic cyst
- Lower lung zone predilection

BHD
- Peripheral, basal dominancy
- Along mediastinum

ILD: LIP, DIP
- Lower lung zone predilection
## Distribution & shape

### Distribution

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<th>Description</th>
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<tr>
<td>Lower dominancy</td>
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<td>Peripheral dominancy</td>
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**No zonal predominancy, Diffuse**
- Uniform shape, sized cysts

**Random distribution**
- Sometimes peripheral, basal, subpleural dominancy
**Distribution & shape**

**Distribution**

- **Upper dominancy**
- **Lower dominancy**
- **No zonal predilection**
- **Peripheral dominancy**

**<Amyloidosis>**
- Predominantly peripheral, associated with nodules

**<BHD>**
- Peripheral, basal dominancy
- Along mediastinum

**<ILD: LIP, DIP>**
- Lower lung zone predilection
Summary
Summary

Cystic lung disease

Step 1: True cysts?
- No
  - Cavitary disease
  - Centrilobular emphysema
  - Bullae
  - Honeycombing
- Yes

Step 2: Localized or diffuse?
- Diffuse
- Localized
  - Incidental cysts
  - Pneumatocele
  - Congenital anomaly
    - CPAM
    - Bronchogenic cyst
- No
  - LAM
  - BHD
  - Malignancy

Step 3: Ancillary feature?
- Yes
- No

Step 4: What kind of feature?
- with nodules
  - PLCH
  - Amyloid
- with GGO
  - LIP
  - DIP
  - PCP

Step 5: Distribution & Shape
**Summary**

**Mimickers**

- **Cavitary disease**
- **Centrilobular emphysema**
- **Bullae**
- **Honeycombing**

- Thick wall (>4mm)  
  Associated with pulmonary consolidation, mass or nodule

- Enlargement and destruction of Alveoli  
  No discrete wall

- Airspace measuring >1cm, Sharply margin, thin wall, associated with emphysema

- Advanced and irreversible fibrotic lung disease  
  3-10mm diameter, clustered, thick wall (1-3mm), subpleural
Summary

Localized cystic lung disease

- Incidental cysts
- Pneumatocele
- Congenital anomaly
  - CPAM
  - Bronchogenic cyst

Aging process
Simple cyst

Transient !!, thin-walled
Gas-filled space

Lower lung dominancy
Type 1 (large cystic lesion), Type 2 (Smaller, uniform cystic lesion)

Commonly filled with fluid, Air-fluid level,
Lower lung dominancy
Summary

Diffuse cystic lung disease without associated feature

- LAM
- BHD
- Malignancy

Diffuse many cysts
Usually uniform, small

Lower dominancy,
Along mediastinum,
Large, various size

Underlying malignancy
Summary

Diffuse cystic lung disease with associated feature

1) with nodules
- PLCH
- Amyloid

   Nodules -> cyst (Temporal heterogeneity)
   Bizarre shape

2) with GGO
- LIP
- DIP
- PCP

   Centrilobular nodules, Perivascular cyst
   Underlying disease

   Cysts in GGO, basilar&periphery

   Thin-walled cysts (30%) : multiple, upper lobe predominance, variation in size + GGO

   Peripheral cysts associated with nodule
   (variable size, calcification)
Summary

Disorder can be included in > 1 category (ex: PLCH)
Not present with cysts (ex: amyloidosis)

→ Multimodality approach !!
(Clinical history, radiologic finding, pathology)
References

1. Suhail Raoof et al., Cystic lung disease Algorithmic approach, CHEST 2016;150(4):945-965


3. Danielle M. Seaman et al., Diffuse cystic lung disease at high-resolution CT, AJR 2011;196:1305–1311


11. Amie Y. Lee et al., Case 182: pulmonary amyloidosis, Radiology 2012;263:929-932

