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CELEBRATING 55 YEARS
INNER SPACES
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Diffuse Cystic Lung Diseases

These are a group of diseases that present with diffuse cysts in the lungs. This flow chart helps (Fig. 1)

The first step is to confirm that the lucencies are true cysts and rule out bronchiectasis, centrilobular emphysema and honeycombing, which are more common (Fig. 2).

Then we check whether the intervening lung parenchyma is normal, or whether there are additional findings. The typical conditions that occur with normal intervening parenchyma (Fig. 3) are the tumors, pulmonary Langerhans cell histiocytosis (PLCH), lymphangioleiomyomatosis (LAM) and cystic metastases, one genetic condition, folliculin deficiency associated ILD (earlier called Birt-Hogg-Dube – BHD syndrome) and lymphoid interstitial pneumonia, typically occurring in patients with Sjogren's and other connective tissue diseases.

Cysts with additional findings may include cysts with nodules within that usually suggest amyloidosis (Fig. 4) or light chain disease.

Cysts may also occur as part of other interstitial lung diseases such as fibrotic hypersensitivity pneumonitis and sarcoidosis.

Once diffuse cystic lung disease is identified, it is important to characterize the lesions and then thoroughly evaluate for an underlying etiolog.

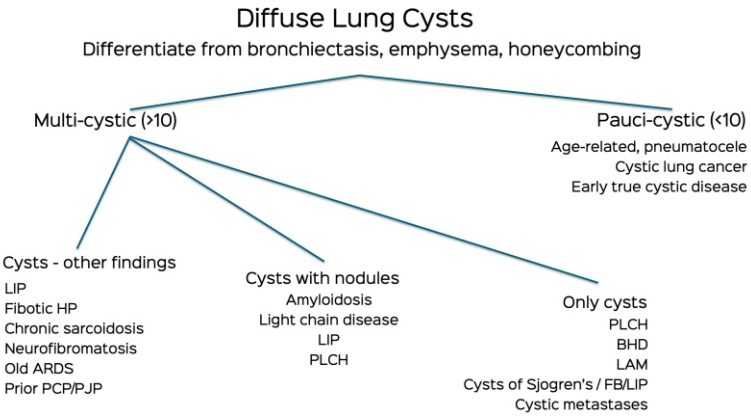

Fig. 1: Flow-chart of diffuse cystic lung disease approach.

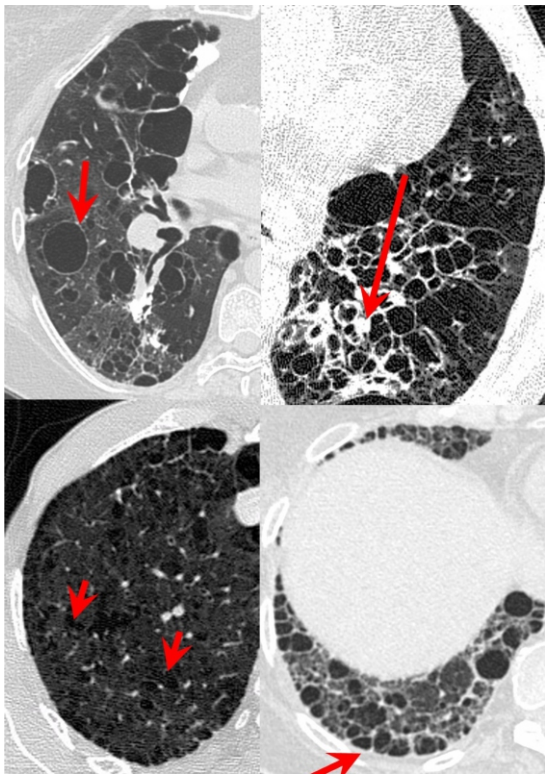

Fig. 2: Cystic lung disease (top left) vs bronchiectasis (top right), centrilobular emphysema (bottom left) and honeycombing (bottom right).

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At a glance

- Diffuse cystic lung disease involves the presence of more than 10 cysts in the lungs
- They are further subclassified based on whether the intervening lung parenchyma is normal or not.
- A detailed evaluation is needed to arrive at the exact cause

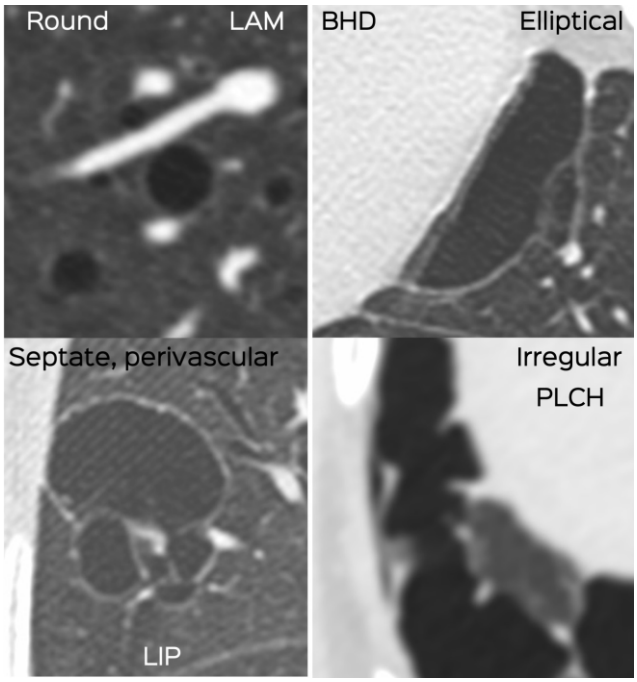

Fig. 3: Cystic diseases with normal intervening parenchyma. Using morphology to differentiate. LAM – lymphangioleiomyomatosis, BHD – Birt-Hogg-Dube syndrome (folliculin deficient ILD), PLCH – pulmonary Langerhans cel

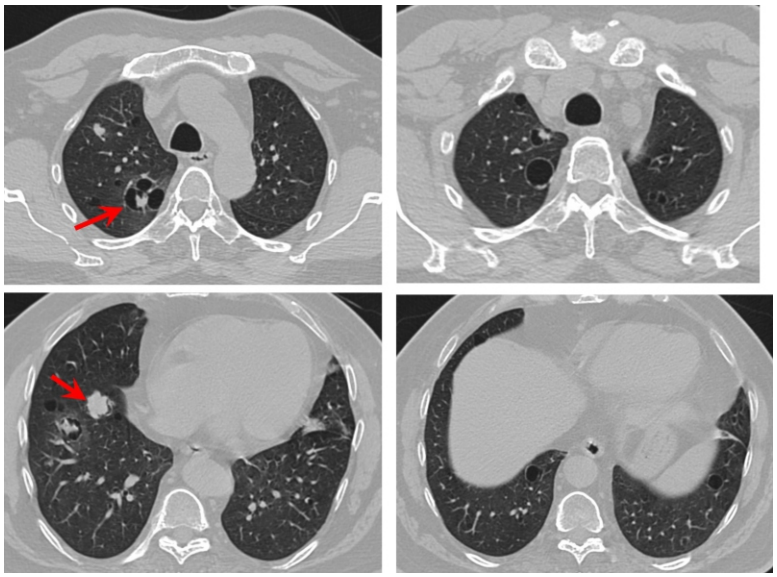

Fig. 4: Cysts with additional findings. Amyloidosis patient with nodules within cysts (arrows).

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