Erdheim-Chester Disease and the lungs

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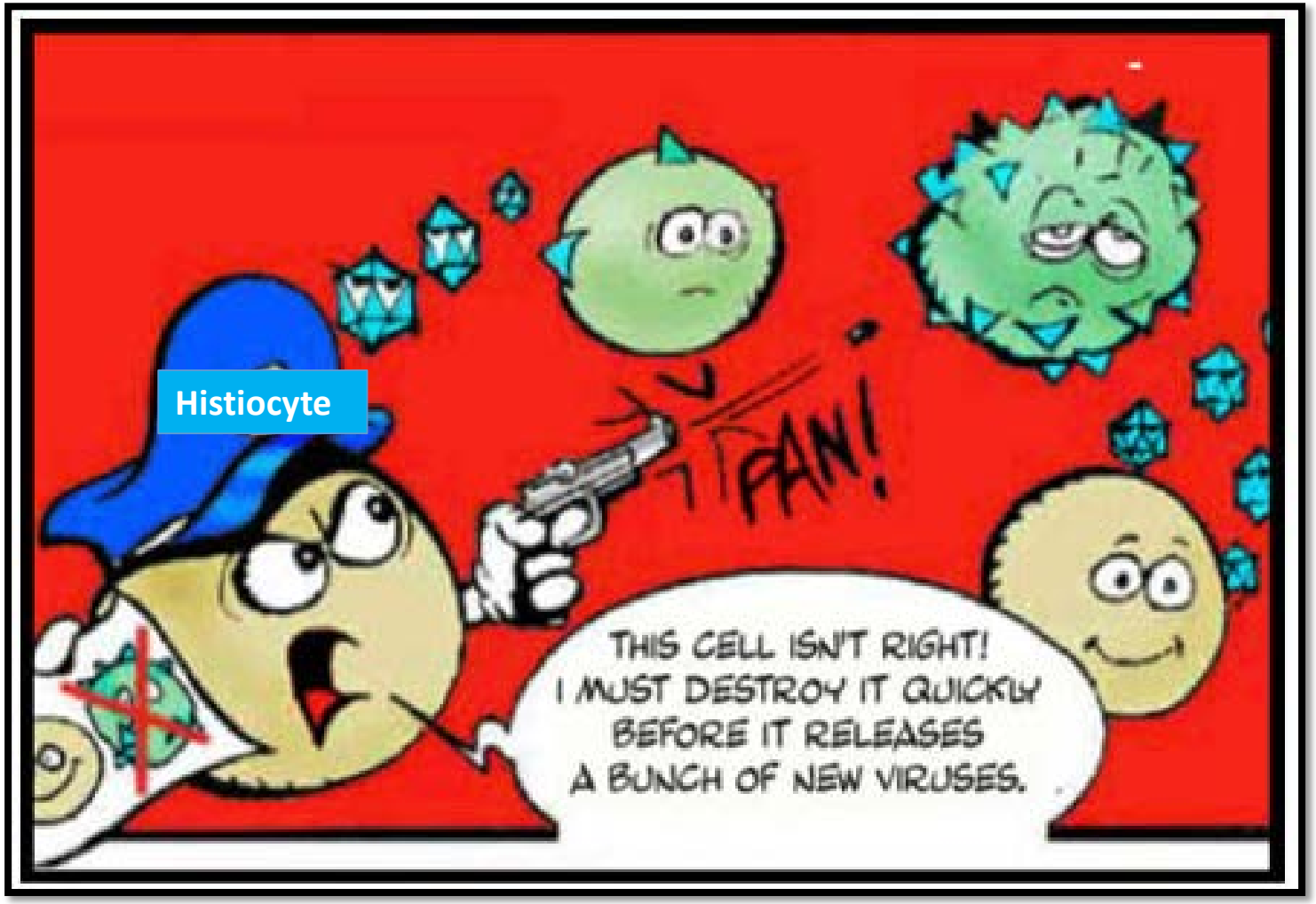
What is the problem?



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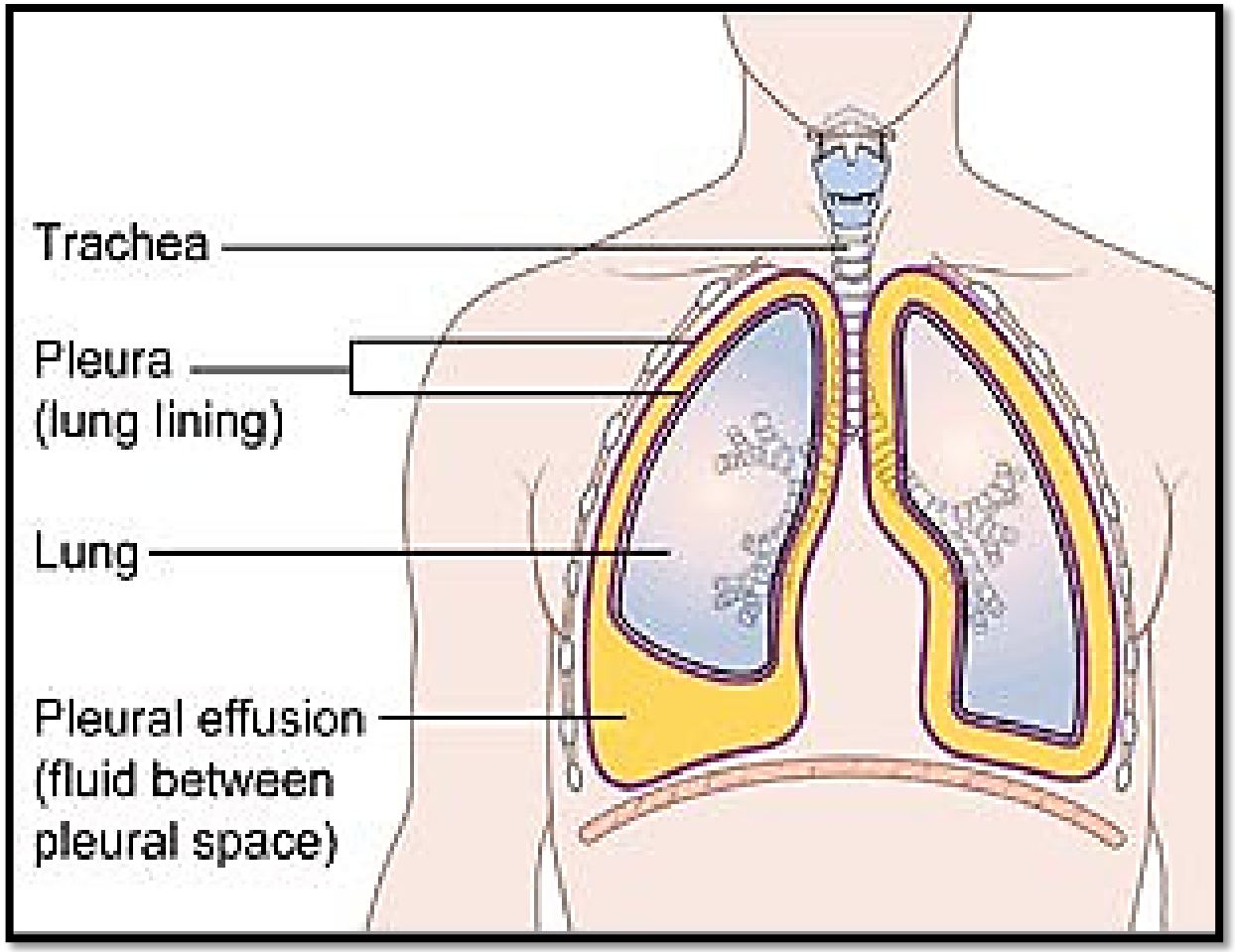
What is the problem?

- Too many histiocytes (cells which normally fight infections)
- Can infiltrate all or some organs including the lungs
- Causes scar-like tissue
- This makes it hard for oxygen to get from the air into the lungs, which can make it hard to breathe

Arnaud L, Pierre I, Beigelman-Aubry C, et al. Pulmonary involvement in Erdheim-Chester disease: a single-center study of thirty-four patients and a review of the literature. Arthritis and rheumatism 2010;62:3504-12.

How many patients have lung involvement?

- One-quarter to one-half of patients will have pulmonary involvement
    - Pleural involvement
    - Parenchymal involvement



What are the symptoms ?

- Cough
- Shortness of breath
- Fatigue
- Chest pain

What testing do I need?

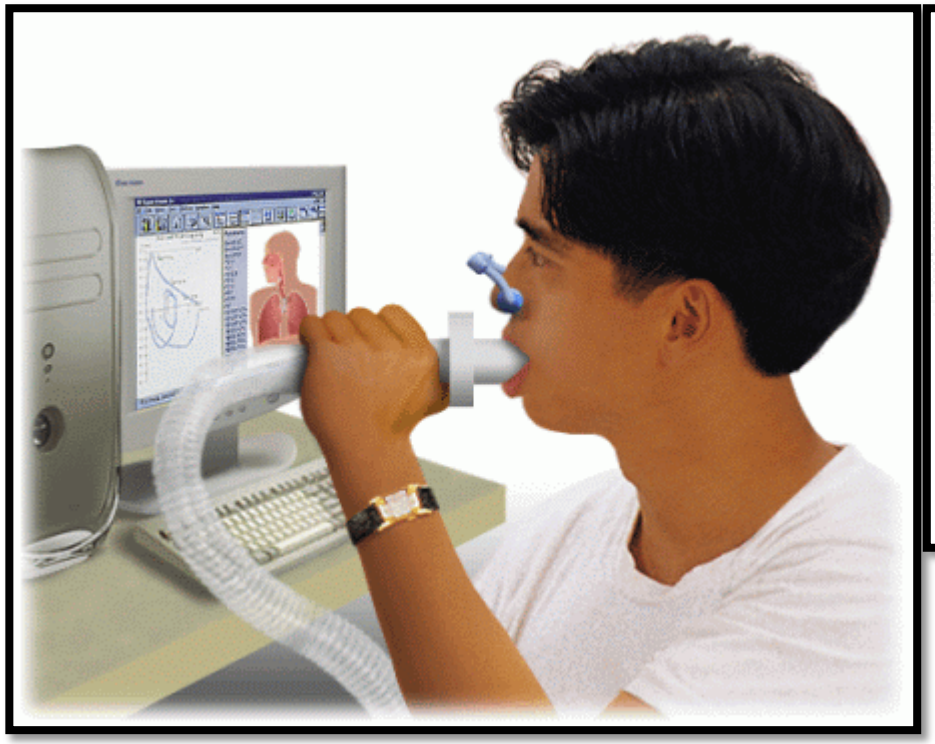
- Pulmonary function test
- 6 minute walk test
- Imaging
- Bronchoscopy

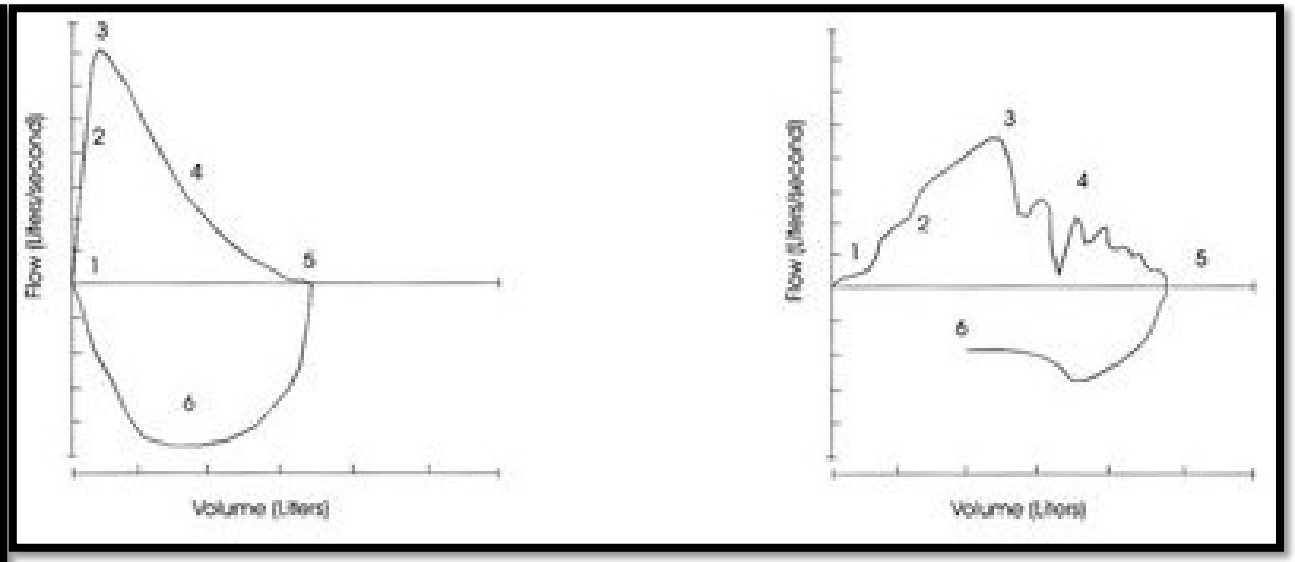
Pulmonary function test (breathing test)

- Pulmonary function testing measures how well you are breathing
- A Complete Pulmonary Function Test often takes 1 ½ hours to complete and can include:
  - Spirometry
  - Lung volumes measurements
  - Diffusing capacity

Pulmonary function test-spirometry

- It involves breathing in as deeply as you can, and then breathing out as hard and as fast as you can into a tube





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Pulmonary function test-lung volumes

- This test measures how much air is in your lungs. In ECD there is decrease in the amount of air in your lungs, also called restriction

- The test involves sitting in a large glass box that looks like a phone booth. While you are in this box, you will breathe in and out through a tube

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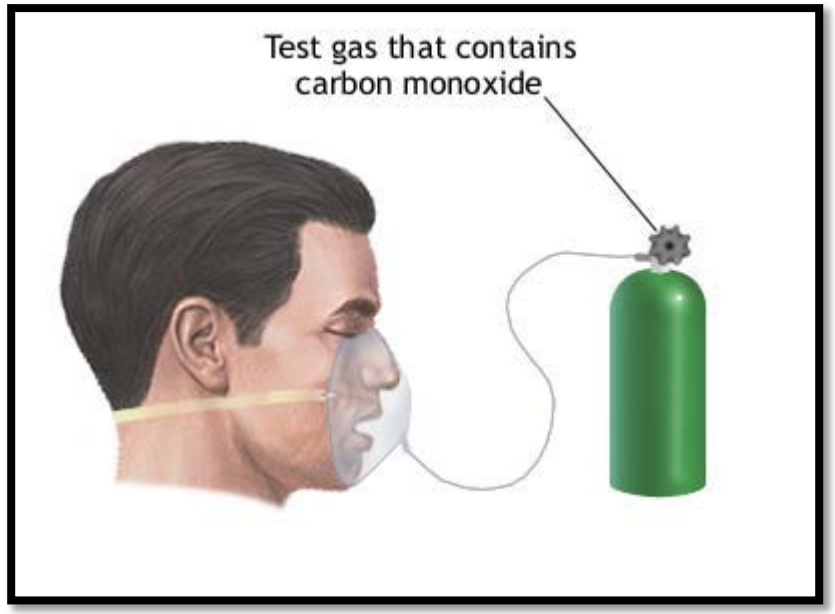
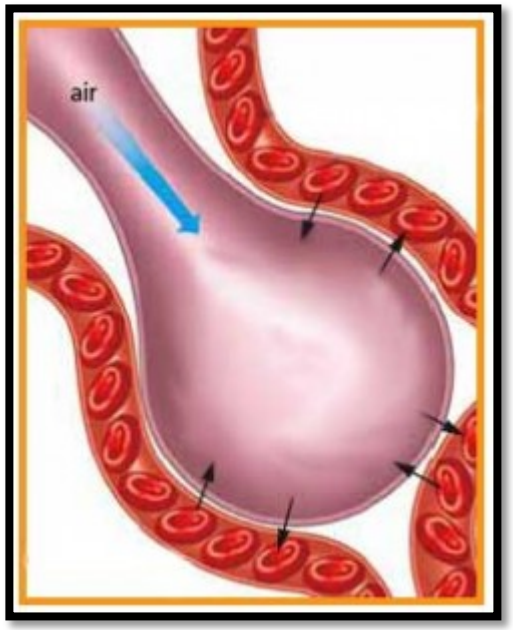
Pulmonary function test-diffusing capacity

- This test measures how well oxygen gets from your lungs into your blood. It involves breathing in a certain gas, and then breathing out into a tube.





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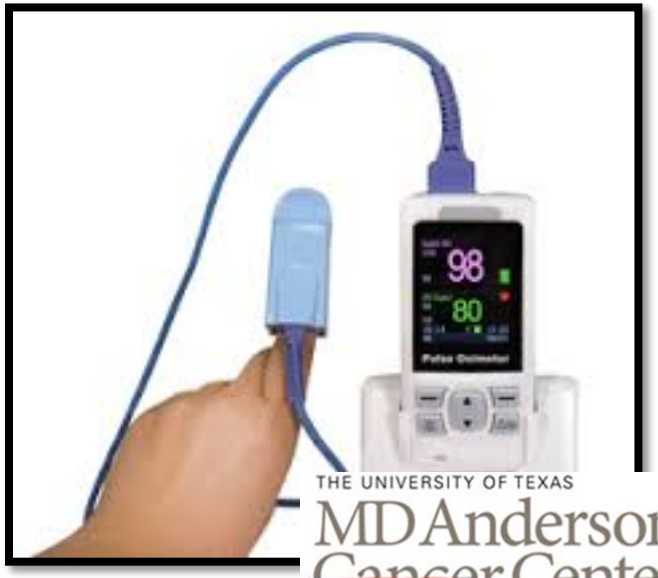
6 minute walk test

- This test measures how far you can walk in 6 minutes. It also measures how much oxygen is in your blood before and after you walk for 6 minutes.
- While you walk, you will wear a sensor on your finger that measures how much oxygen is in your blood
- Healthy subjects can typically walk 400 to 700 m



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Imaging

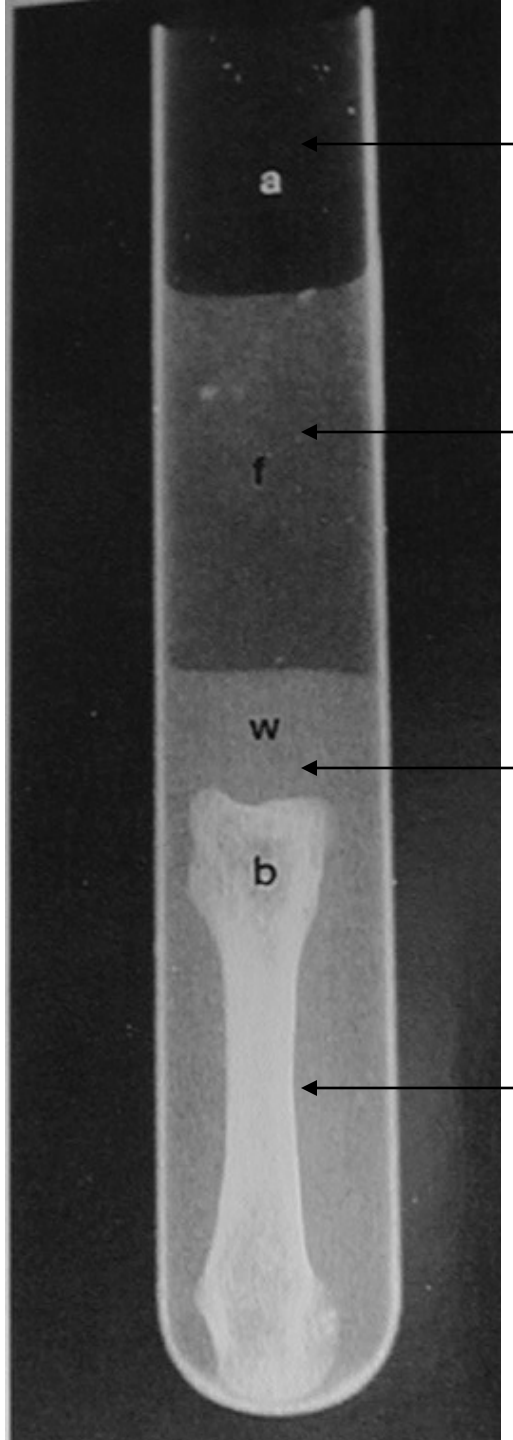
- Chest x-ray
- Computerized tomography (CT) scan

Chest X-ray

- The lung damage associated with ECD disease often shows up in characteristic patterns on chest X-rays

- Occasionally, the chest X-ray is normal

Glass
Test Tube



Air

Fat

Water

Bone
+
Water

Metal

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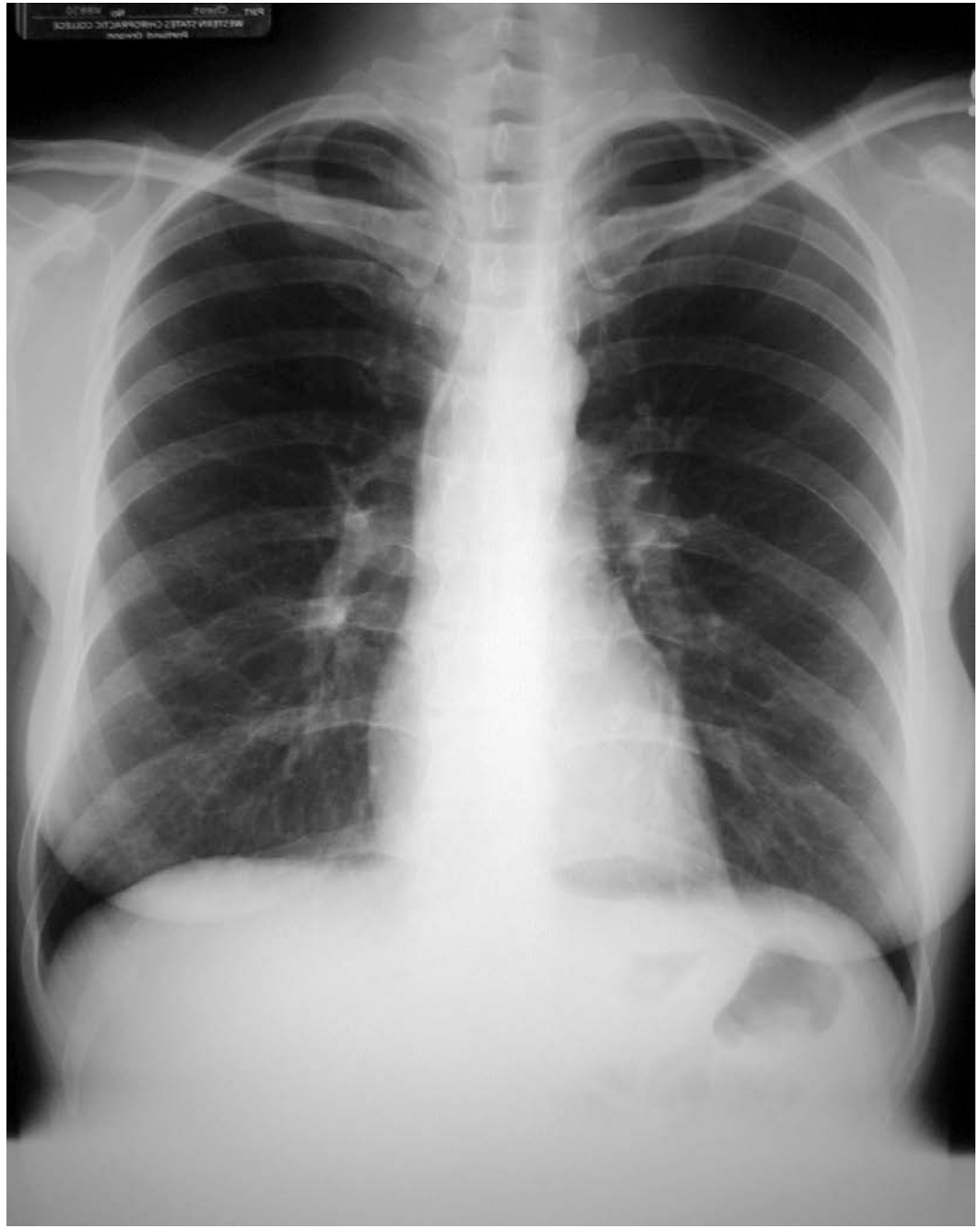

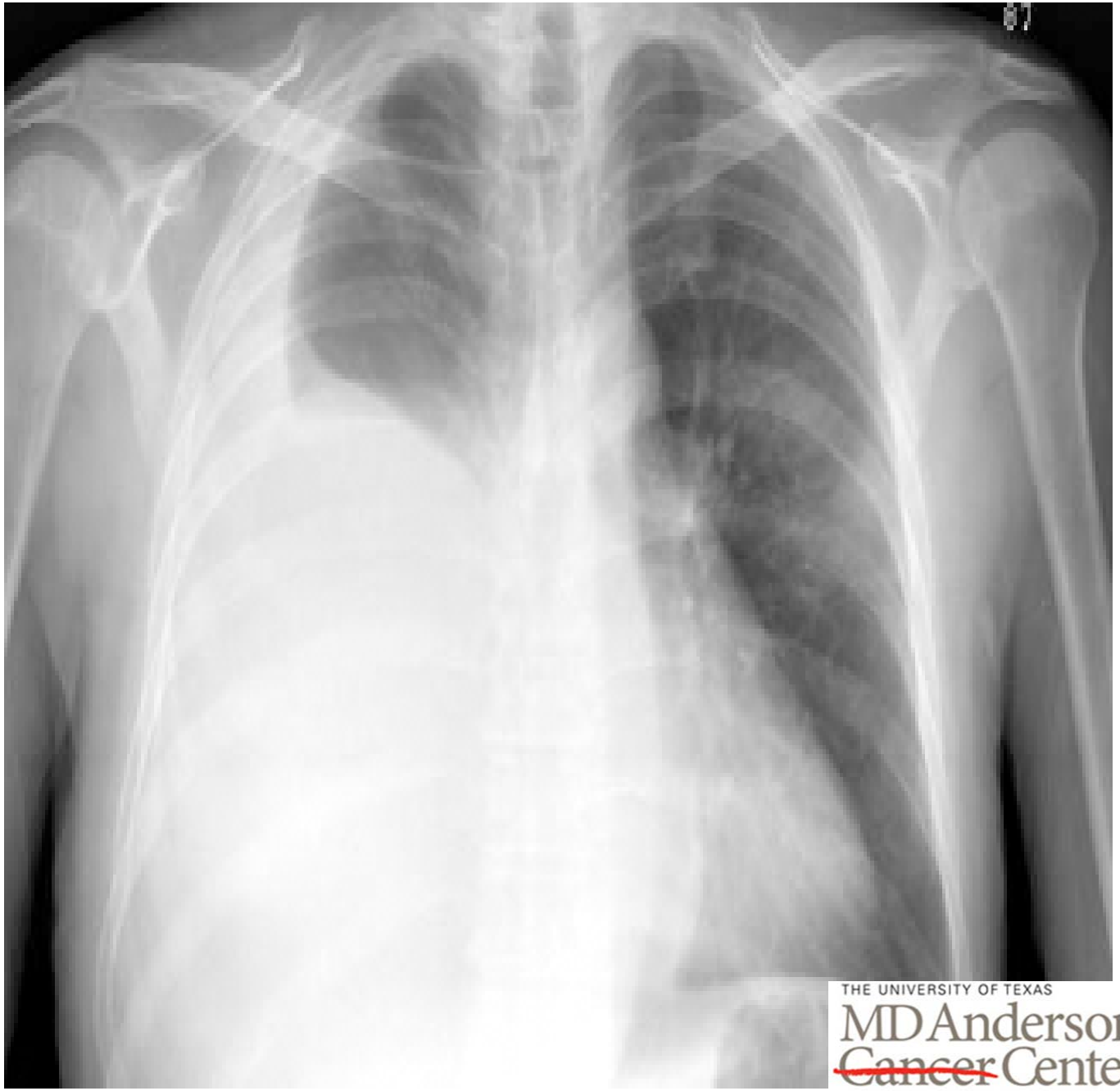

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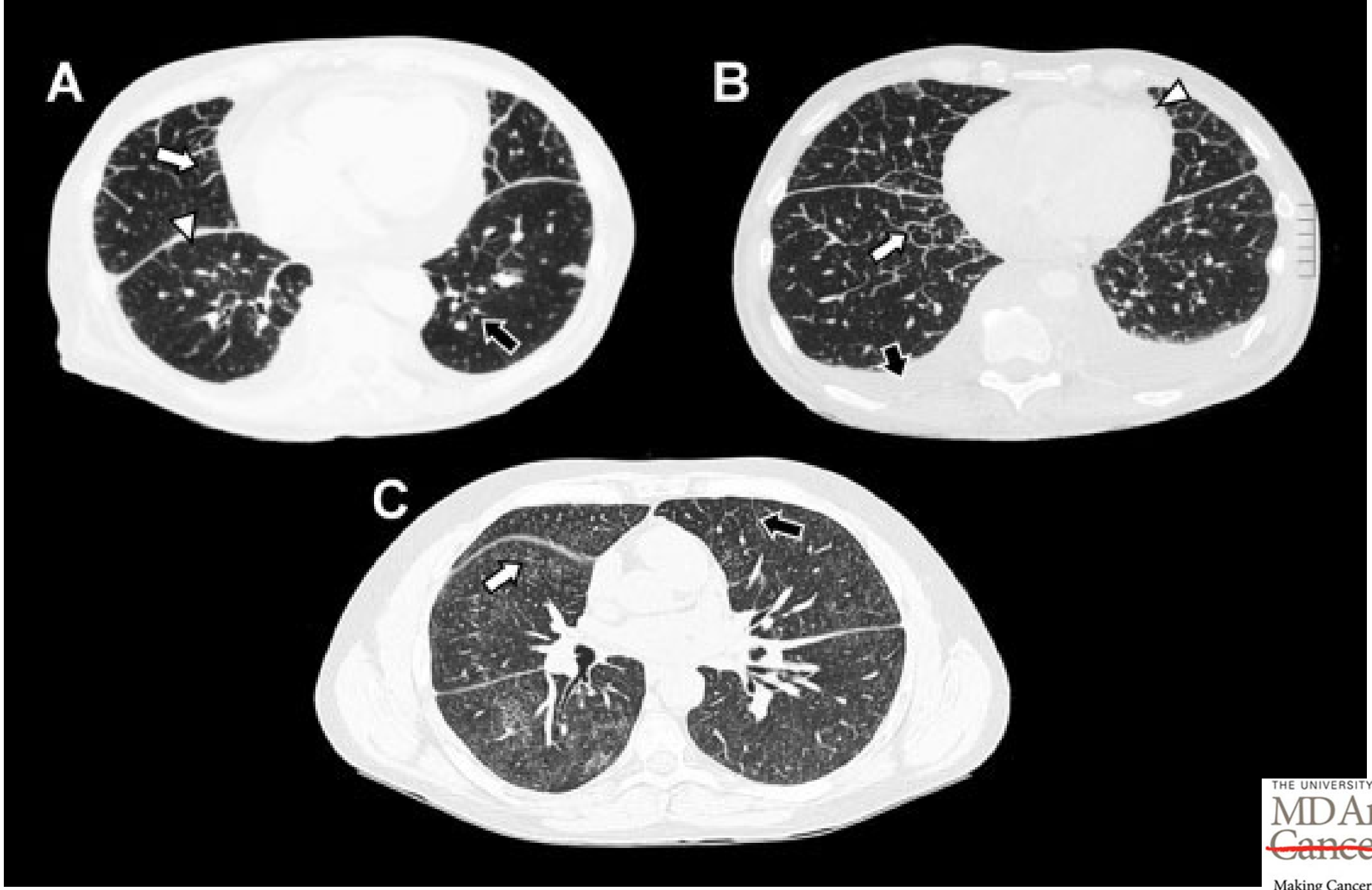
Computerized tomography (CT) scan

- CT scanners use a computer to combine X-ray images taken from many different angles to produce cross-sectional images of internal structures



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Wittenberg KH, Swensen SJ, Myers JL. Pulmonary involvement with Erdheim-Chester disease: radiographic and CT findings. AJR American journal of roentgenology 2000;174:1327-31.

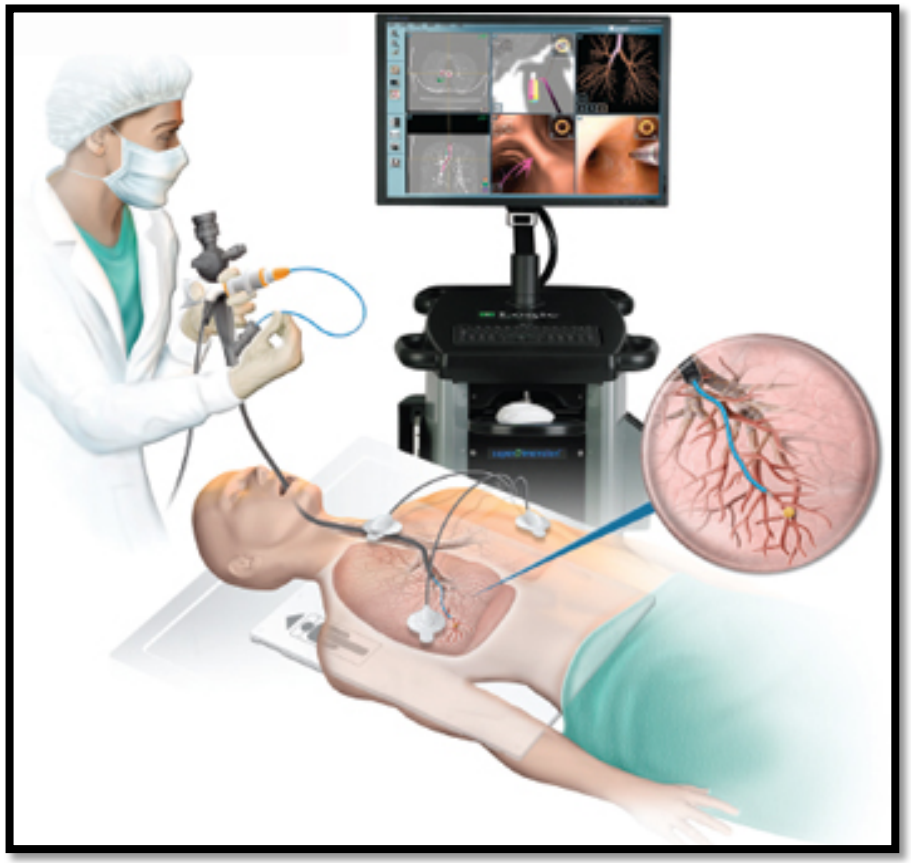
Bronchoscopy

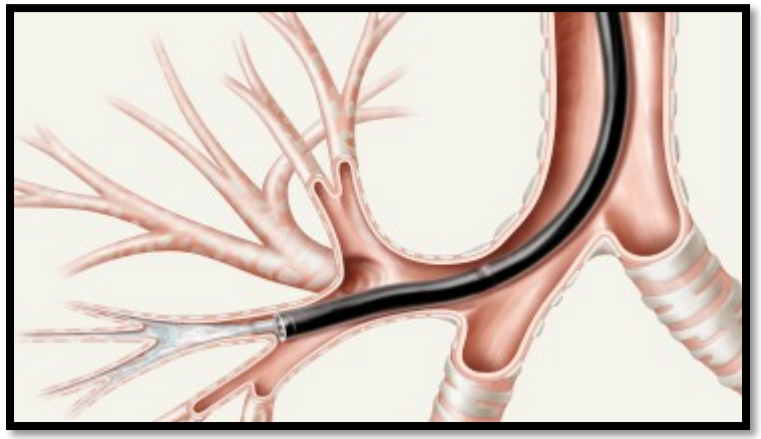
- In this procedure, we remove fluid or a very small tissue sample — generally no larger than the head of a pin — using a small, flexible tube (bronchoscope) that's passed through your mouth or nose into your lungs

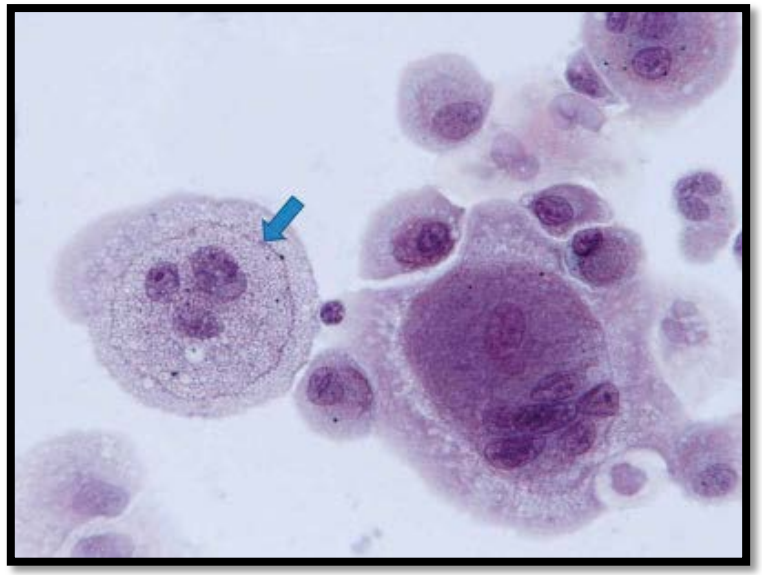
- The serious risks of bronchoscopic biopsy include bleeding or a deflated lung, which may require treatment

- More common side effects are temporary sore throat and hoarseness

Bronchoscopy with bronchoalveolar lavage







Double-contoured histiocytes in the bronchoalveolar lavage
fluid of a patient with Erdheim-Chester disease. (arrow)

Wittenberg KH, Swensen SJ, Myers JL. Pulmonary involvement with Erdheim-Chester disease: radiographic and CT findings. AJR American journal of roentgenology 2000;174:1327-31.

Diagnosis

- Pulmonary ECD is considered highly probable when
  - Presence of radiological hallmarks of interstitial lung disease in ECD patients
  - The detection of CD68(+), CD1a(−) histiocytes in bronchoalveolar lavage fluid confirms the diagnosis of pulmonary ECD

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Treatment

- Not all patients with ECD disease require treatment
- Treatment varies with most patients undergoing several kinds of treatments
- Pulmonary involvement is not the main indication for therapy
- Symptoms due to pleural effusion, low oxygen or cough may need symptomatic treatment

Pleural effusion-fluid around the lungs

- Draining pleural effusion-Thoracentesis

Pleural effusion-fluid around the lungs

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Low oxygen

- Supplemental oxygen
- When oxygen is bellow 88-89%

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Oxygen therapy

- Using oxygen can't stop lung damage, but it can:
- Make breathing and exercise easier
- Prevent or lessen complications from low blood oxygen levels
- Reduce blood pressure in the right side of your heart
- Improve your sleep and sense of well-being
- You're most likely to receive oxygen when you sleep or exercise, although some people may use it round-the-clock

Cough

- Cough suppressants
  - Over the counter
  - Prescription

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Prognosis

- The researchers concluded that pulmonary involvement of ECD can add to the morbidity the disease but has limited impact on the overall prognosis of the disease

Important !

Eat well

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Remain active

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NO smoking !!!!!

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Thank you