

THE INTERNATIONAL
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OUR MYOSITIS HEROES

SEPTEMBER 7-10, 2023

SHERATON SAN DIEGO HOTEL & MARINA



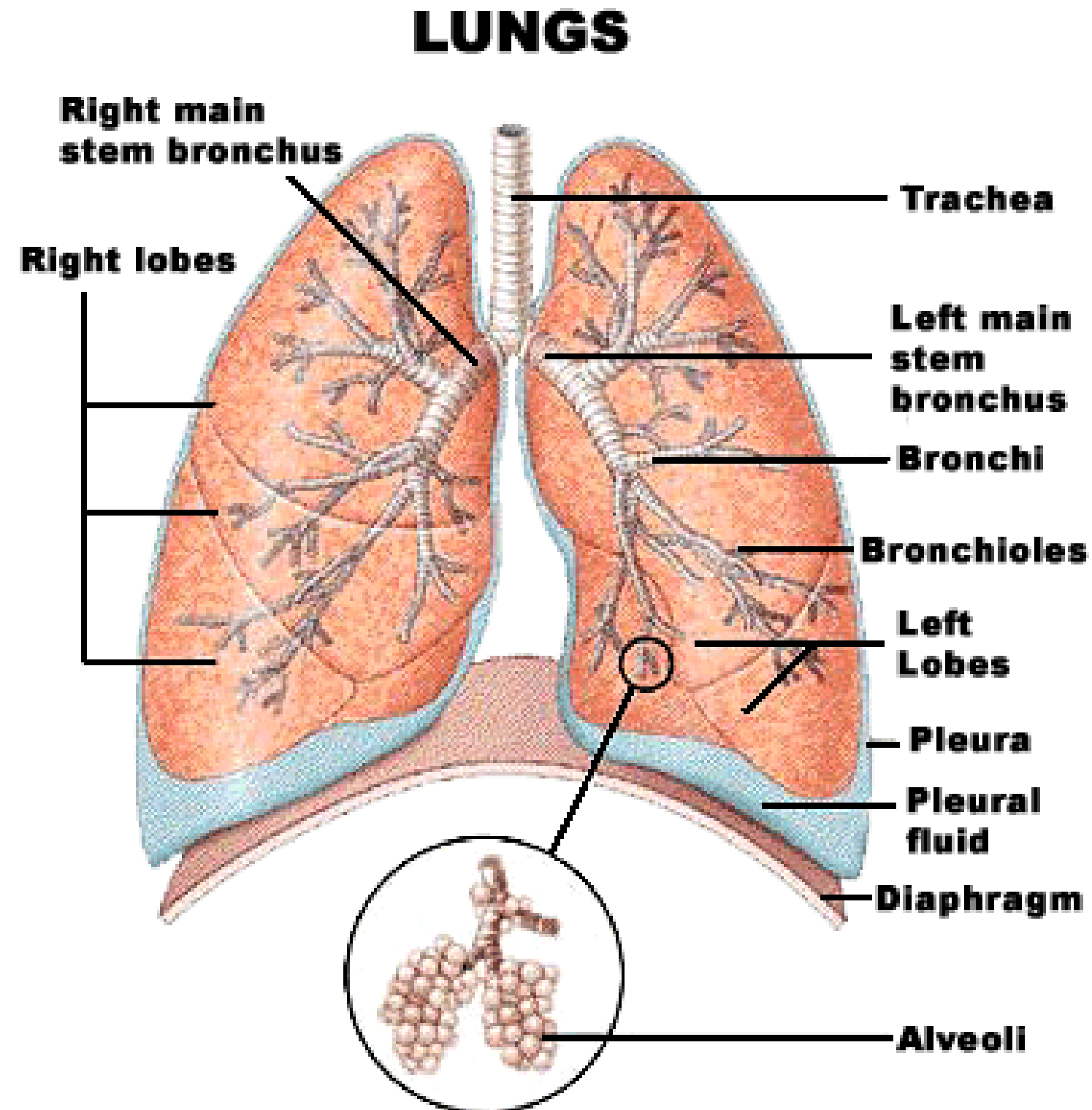
ILD: What do all these tests mean?

Sonye K. Danoff, MD, PhD

Professor of Medicine

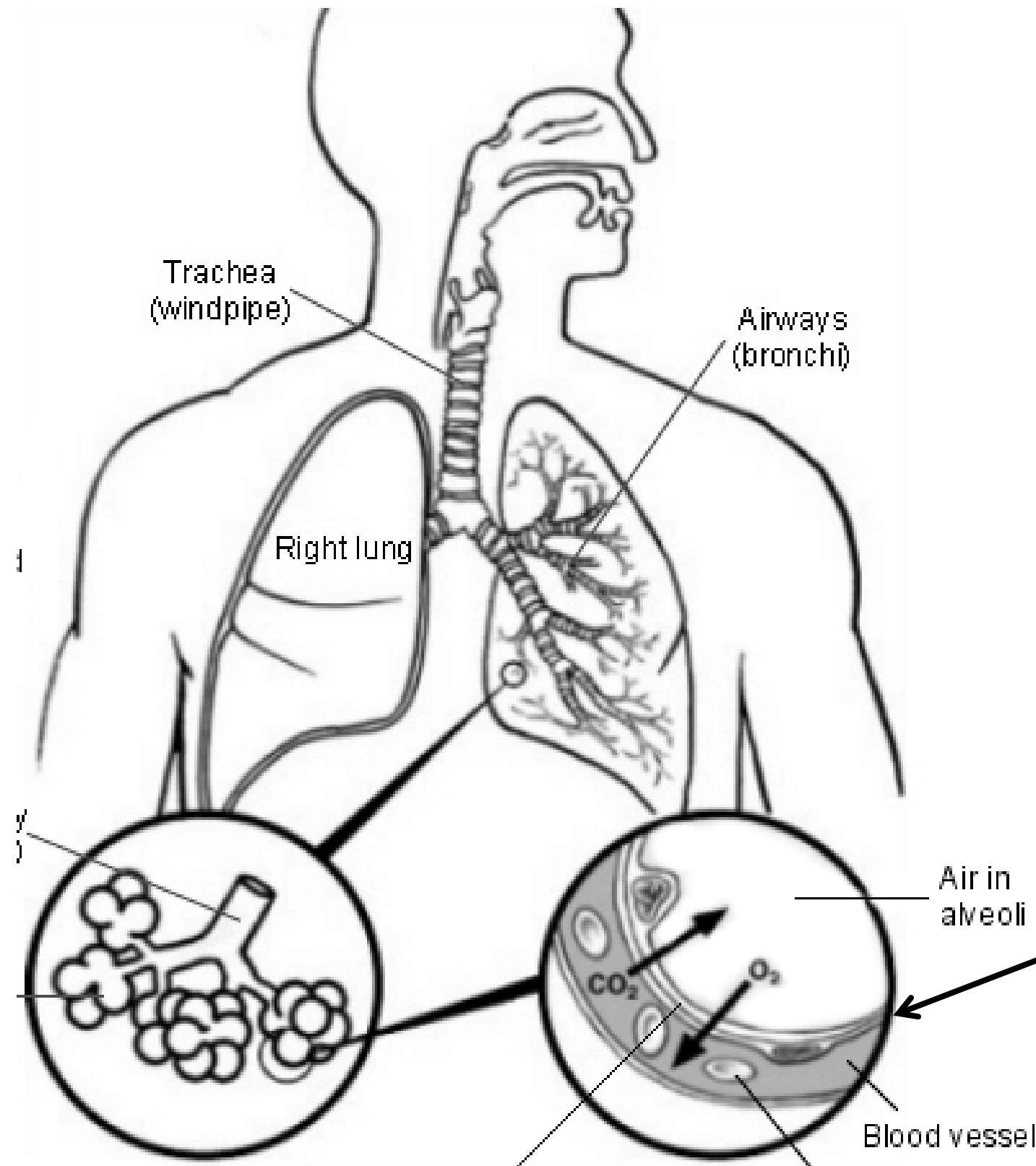
Director, Johns Hopkins ILD/PF Program

1) What makes up the lung?



- Airways (tubes that connect the lung to the mouth)
- Blood vessels (carry the blood cells)
- Alveoli (air sacs)

2) What happens in healthy lungs?



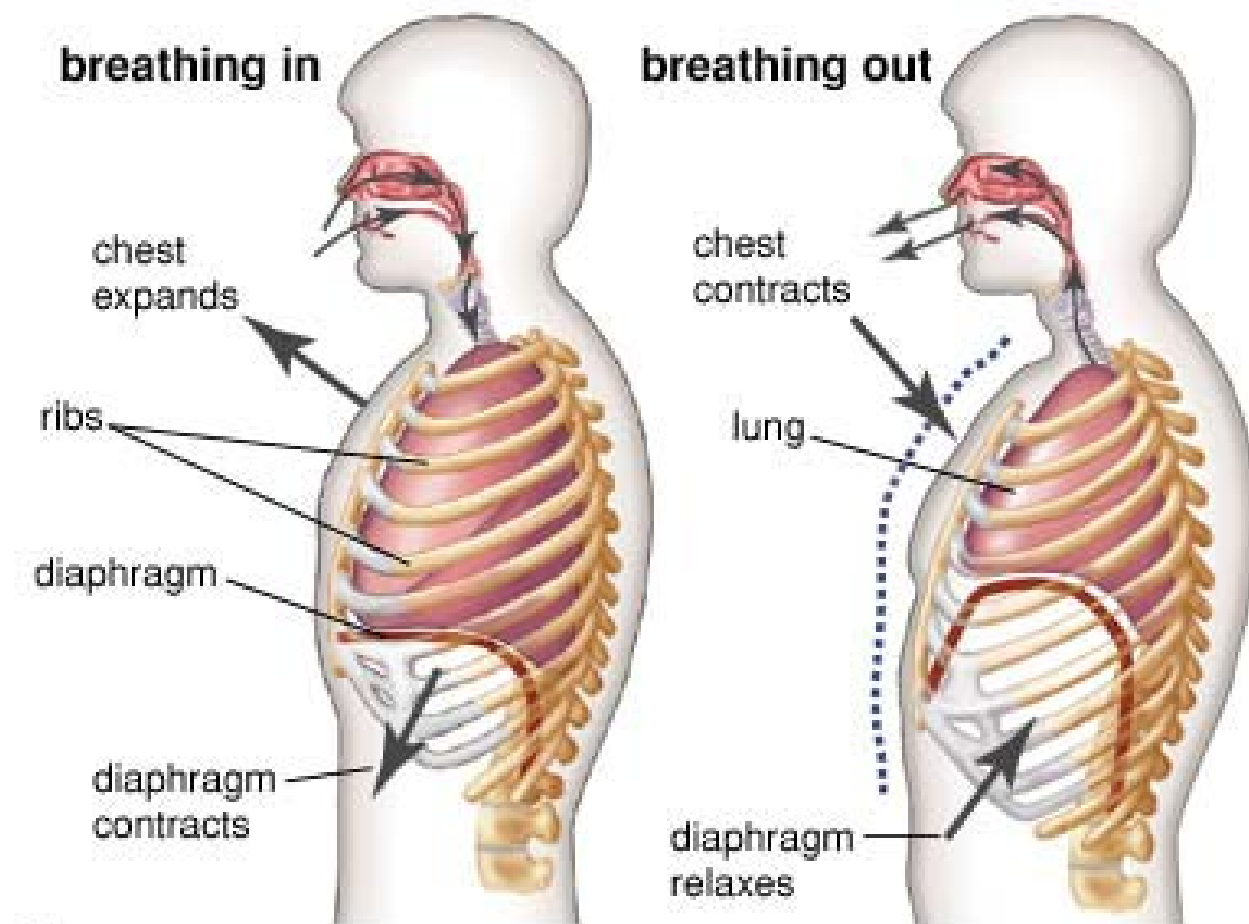
Air enters the mouth and passes down the trachea and airways into the air sacs or alveoli.

Air contains a mixture of gases including oxygen.

Oxygen is absorbed in the alveoli where the air in the lung comes close to the red blood cells in the blood vessels.

3) Why do people with “myo”sitis get interstitial lung disease?

- Does the lung have muscles? Yes & No
- Muscles of respiration
 - Diaphragm
 - Intercostal muscles
- Lung is also injured
 - Even though it isn't a muscle



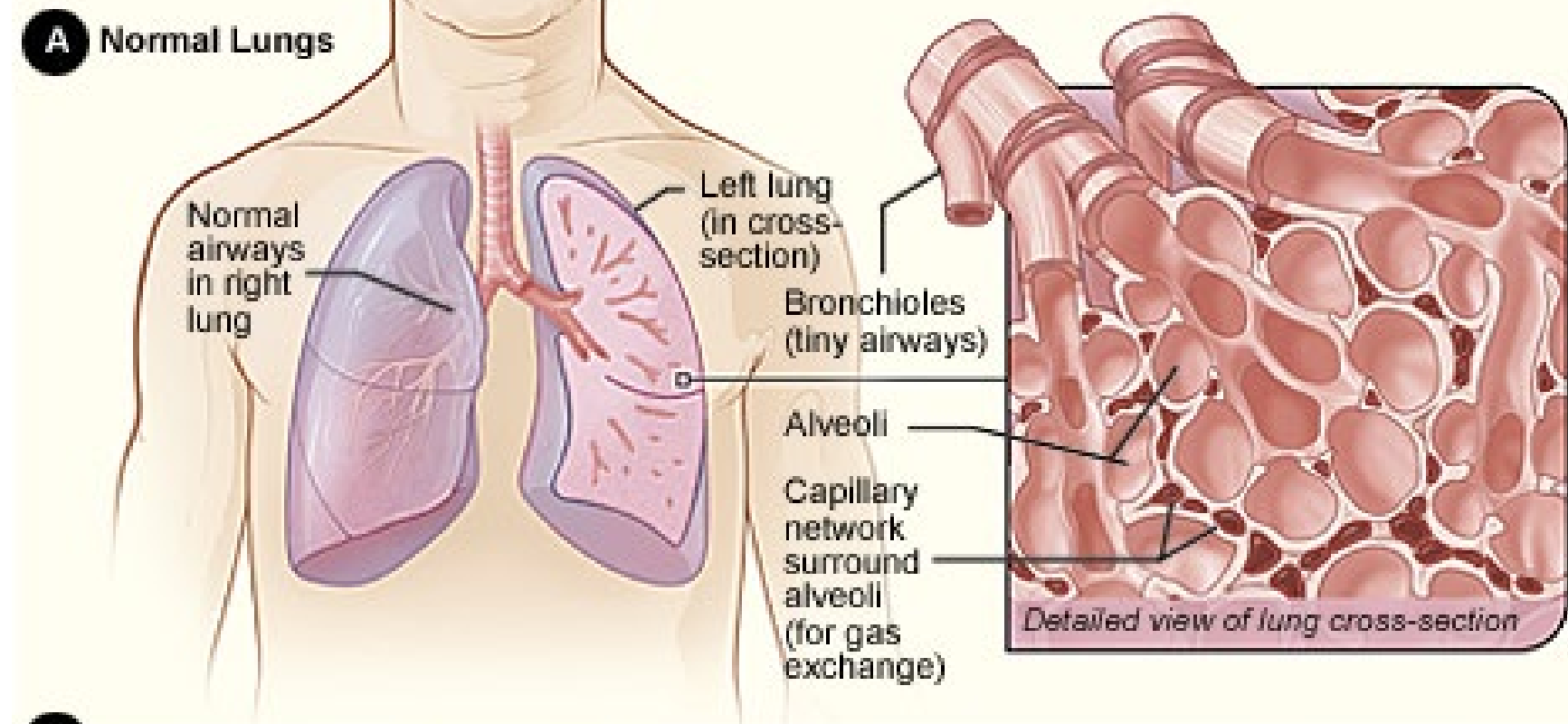
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4) What is interstitial lung disease?

- Injury to the lung that happens when immune cells are activated (inflammation)
- Can result in scarring (fibrosis) in some cases

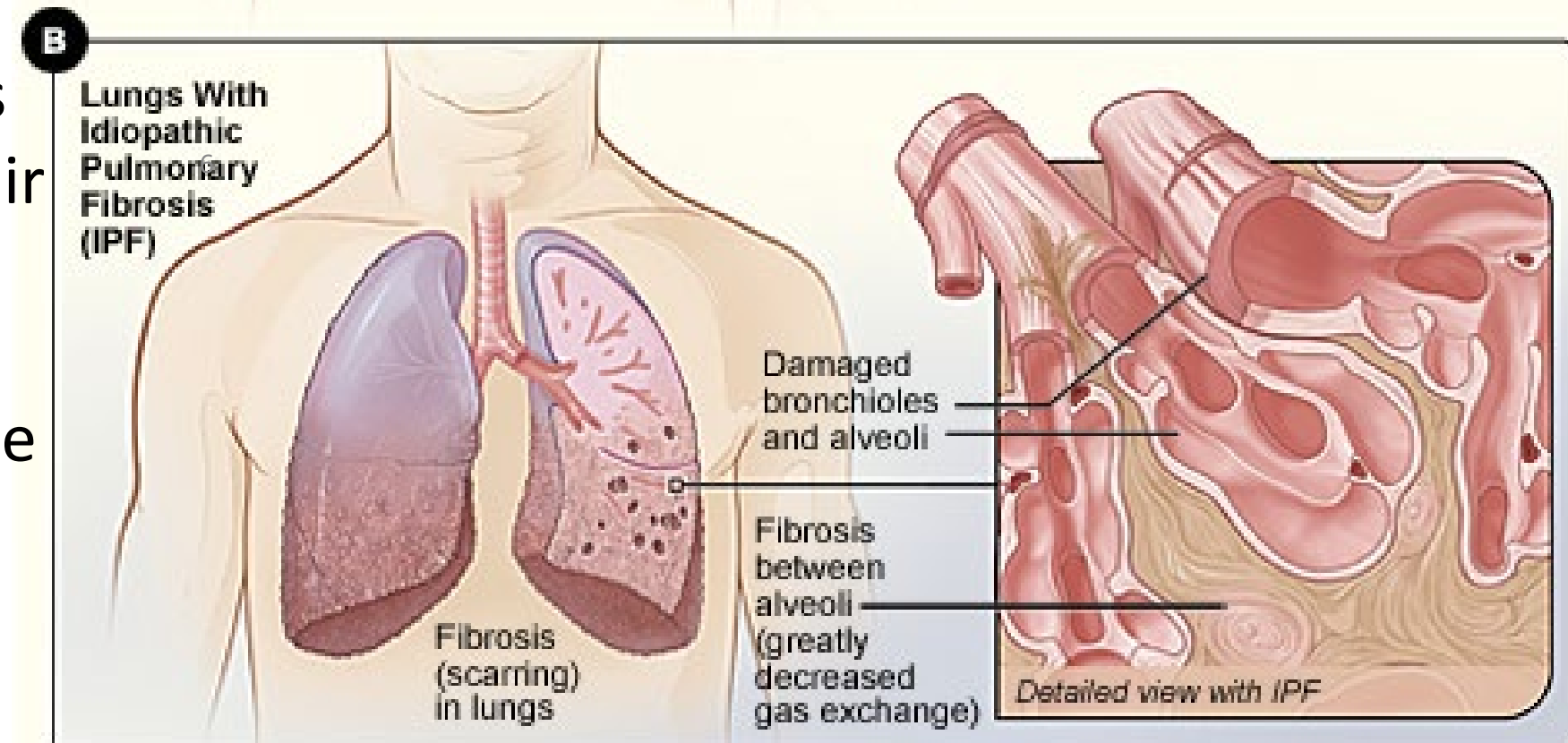


5) What is the effect of interstitial lung disease?



Inflammation and scarring blocks the transfer of oxygen from the air into the blood.

Oxygen is needed in all cells in the body in order to burn fuel efficiently.



6) What are the symptoms of interstitial lung disease?

- Each person is different
- Some symptoms are common
 - Shortness of breath
 - especially with activity
 - Cough
 - Feeling tired



7) How is Interstitial lung disease diagnosed?

- Velcro-like “Crackles” heard through a stethoscope
- Chest xrays and CT scans
- Breathing tests called Pulmonary Function Tests (PFTs)
- Lung biopsies are *rarely* needed
- Other tests: echo, labs



Chest CT Scan



Normal Lung



Interstitial Lung Disease

Pulmonary Function Tests:

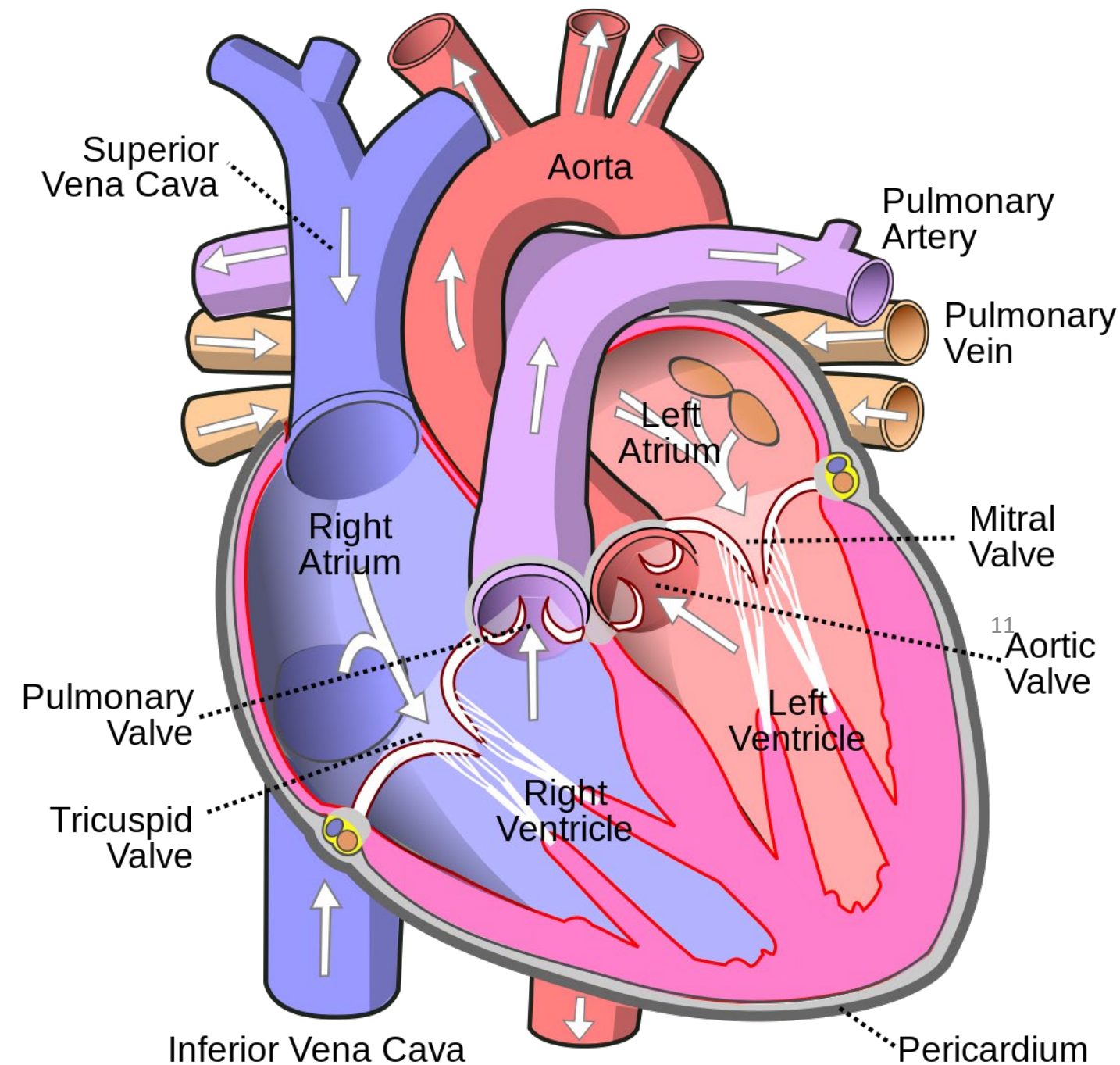
Size of the Lung

Ability to absorb
oxygen

	<u>Pred</u>	<u>Actual</u>	<u>%Pred</u>
---- SPIROMETRY ----			
→ FVC (L)	3.12	2.50	80
FEV1 (L)	2.44	2.00	81
FEV1/FVC (%)	79	80	100
FEF 25-75% (L/sec)	2.15	1.81	84
FEF 50% (L/sec)	3.53	2.45	69
FEF Max (L/sec)	6.17	6.43	104
---- LUNG VOLUMES ----			
TGV (L)	2.92	2.13	72
SVC (L)	3.12	2.81	90
→ TLC (Pleth) (L)	5.14	4.57	88
RV (Pleth) (L)	2.09	1.75	83
RV/TLC (Pleth) (%)	41	38	93
ERV (L)	0.90	0.28	31
IC (L)	2.22	2.44	109
Raw (cmH2O/L/s)	1.86		
---- DIFFUSION ----			
→ DLCOunc (ml/min/mmHg)	21.46	15.33	71
DLCOcor (ml/min/mmHg)	21.46		
DL/VA (ml/min/mmHg/L)	4.36	3.49	79
VA (L)	5.15	4.40	85

Normal:
≥80%
Predicted

Echocardiograms: looking at the heart with sound waves



- The right side of the heart pumps to the lungs
- The left side of the heart pumps to everything else
- High blood pressure refers to excess pressure on the left side of the heart
- Pulmonary hypertension (PH) refers to excess pressure on the right side of the heart

Labs and more labs

- At diagnosis
 - Antibody testing
 - Basic labs (blood count and chemistry)
 - Tests for other types of ILD
 - Labs for PH
- During treatment
 - Medication safety monitoring (based on which medication is used)
 - Sometimes, more antibody tests
 - Labs for PH

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8) Why is it important to identify interstitial lung disease?

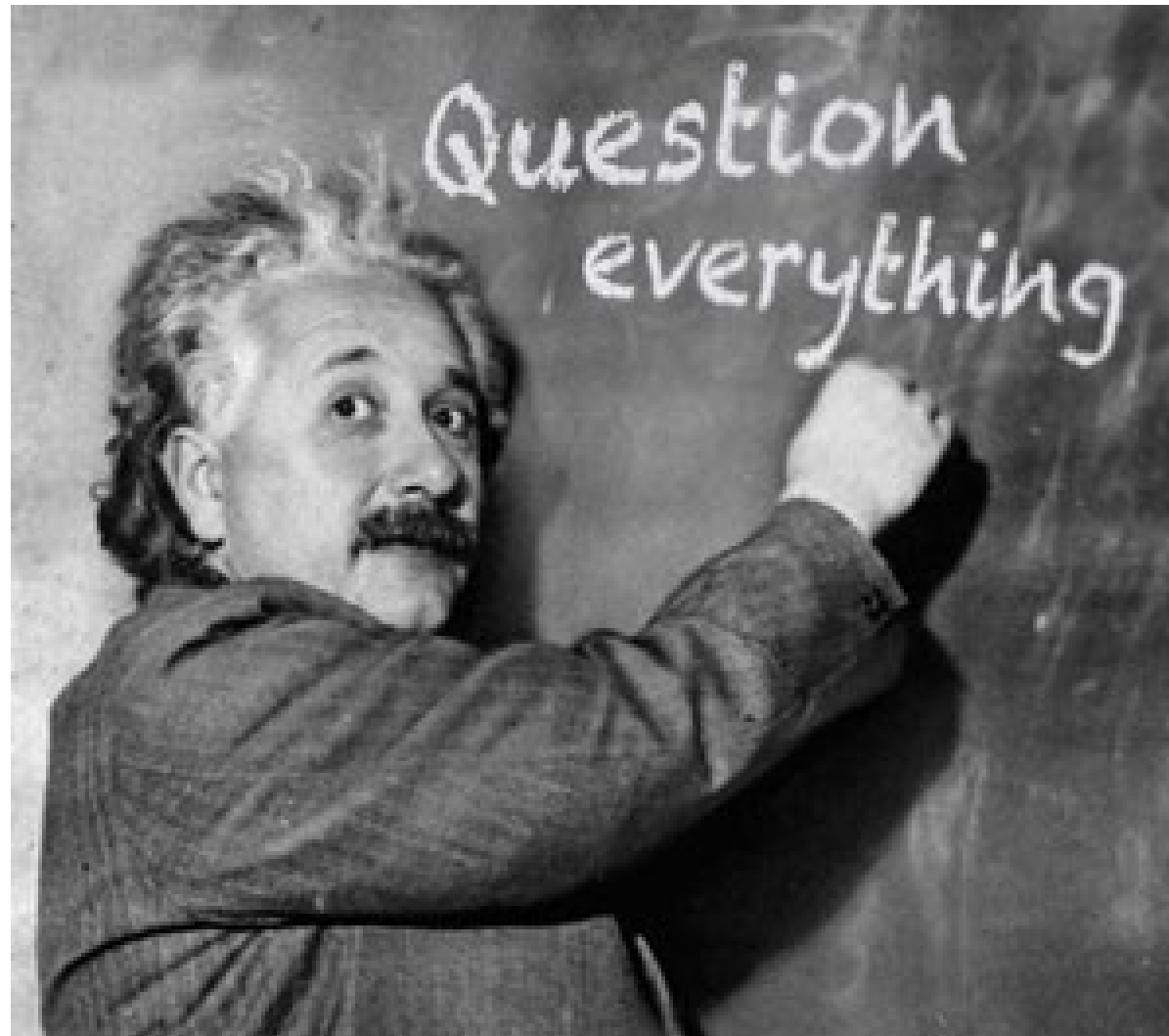
- Symptoms of shortness of breath may be less apparent in patients who are weak
- Interstitial lung disease may worsen even as muscle and skin disease improves
- Treatment decisions should often be driven by the lung disease
- Interstitial lung disease can be life-threatening

9) What could interstitial lung disease mean for my life?



- Different for each person
- Medications often help
- Oxygen
- Pulmonary rehabilitation

10) Questions?





Insert Text



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Thank you!