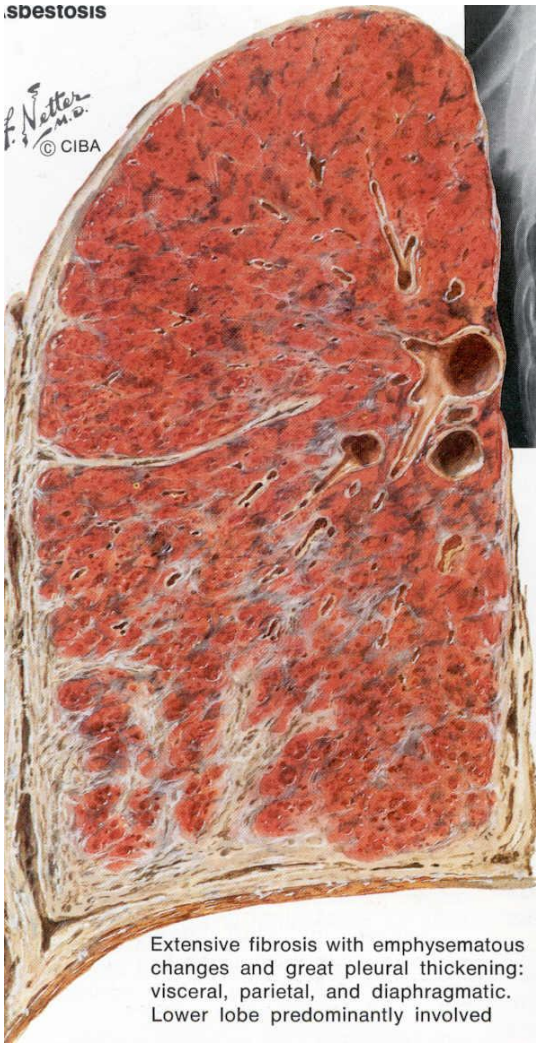
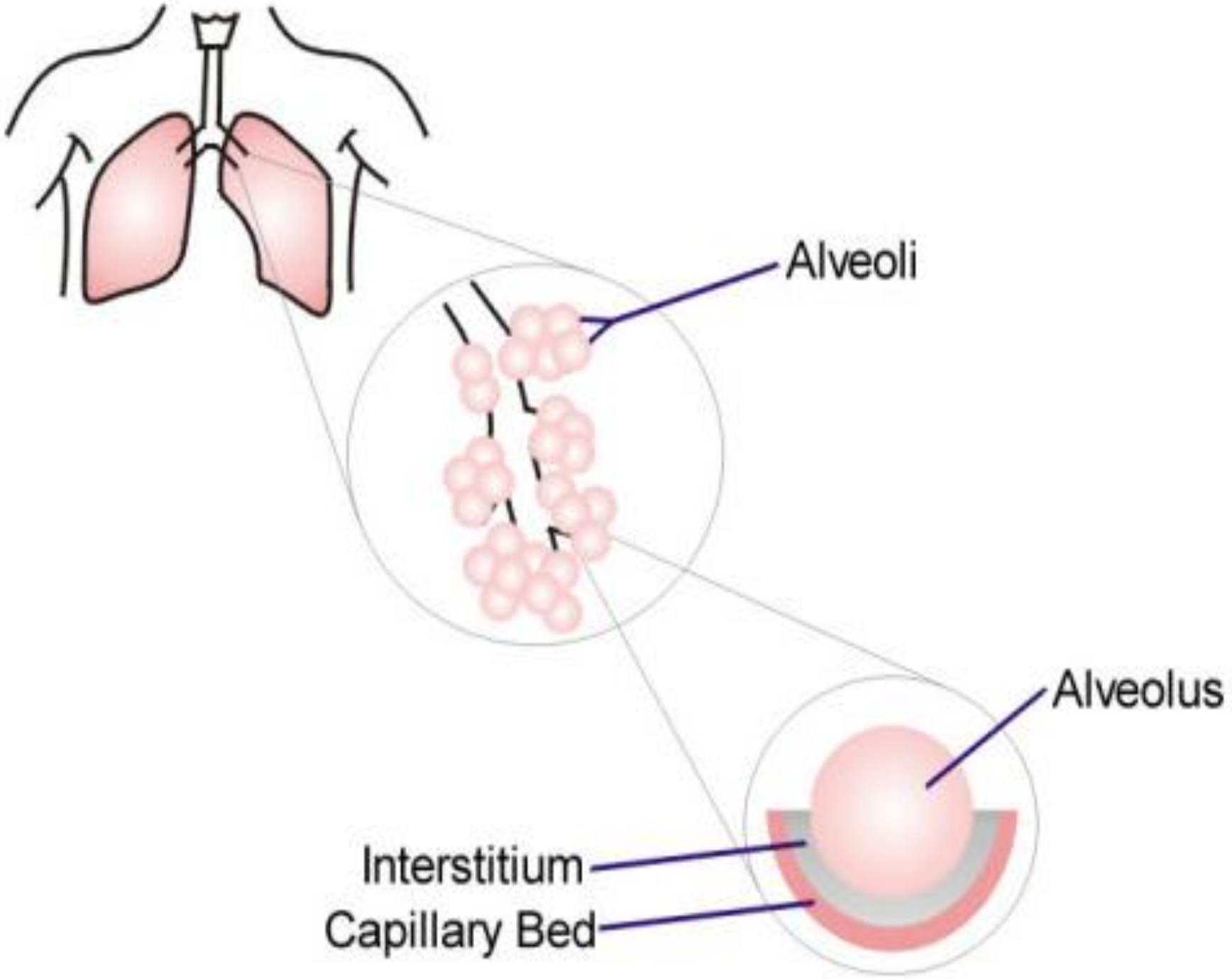
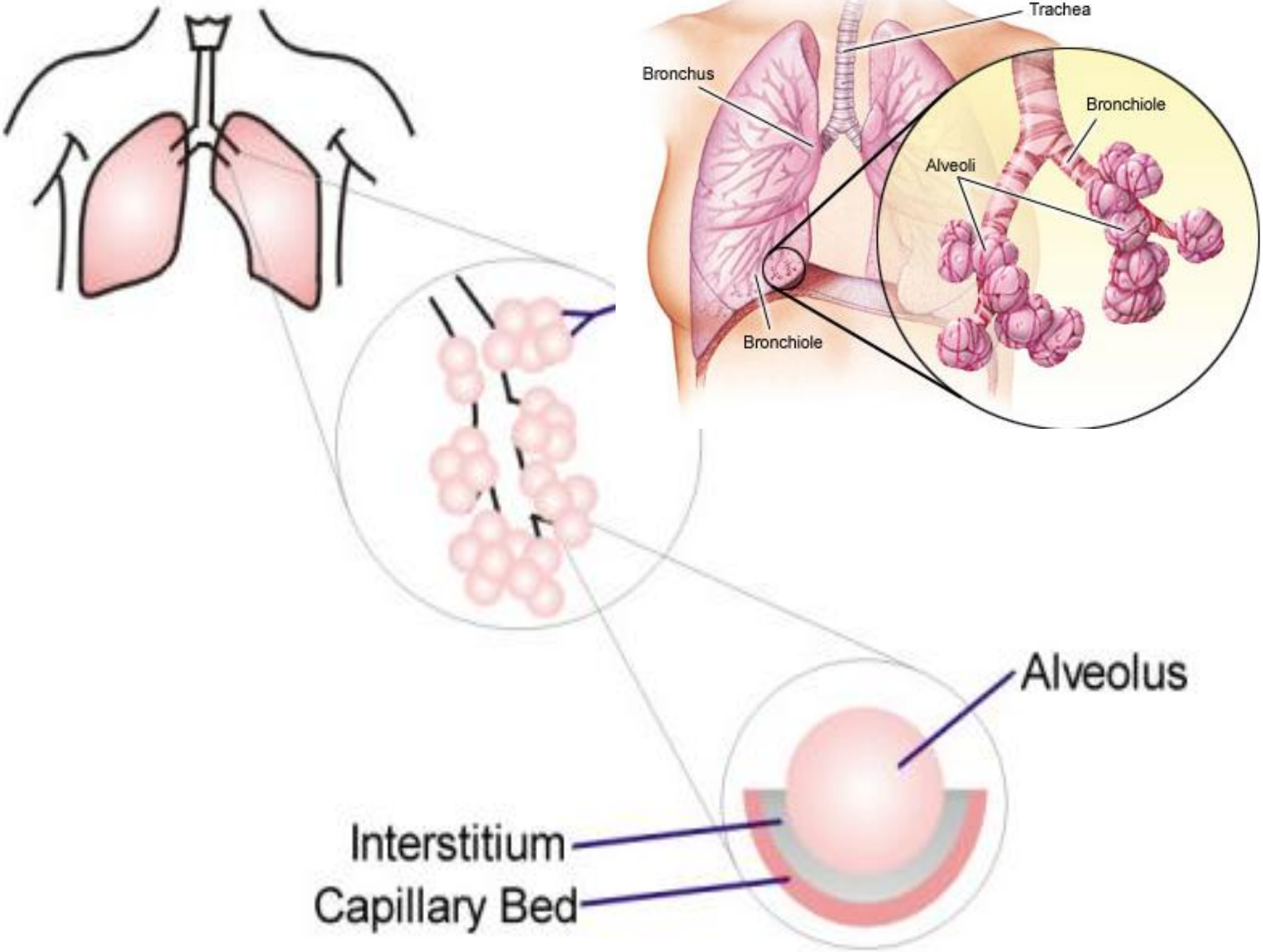
Interstitial lung diseases
Restrictive lung diseases
Lung fibrosis

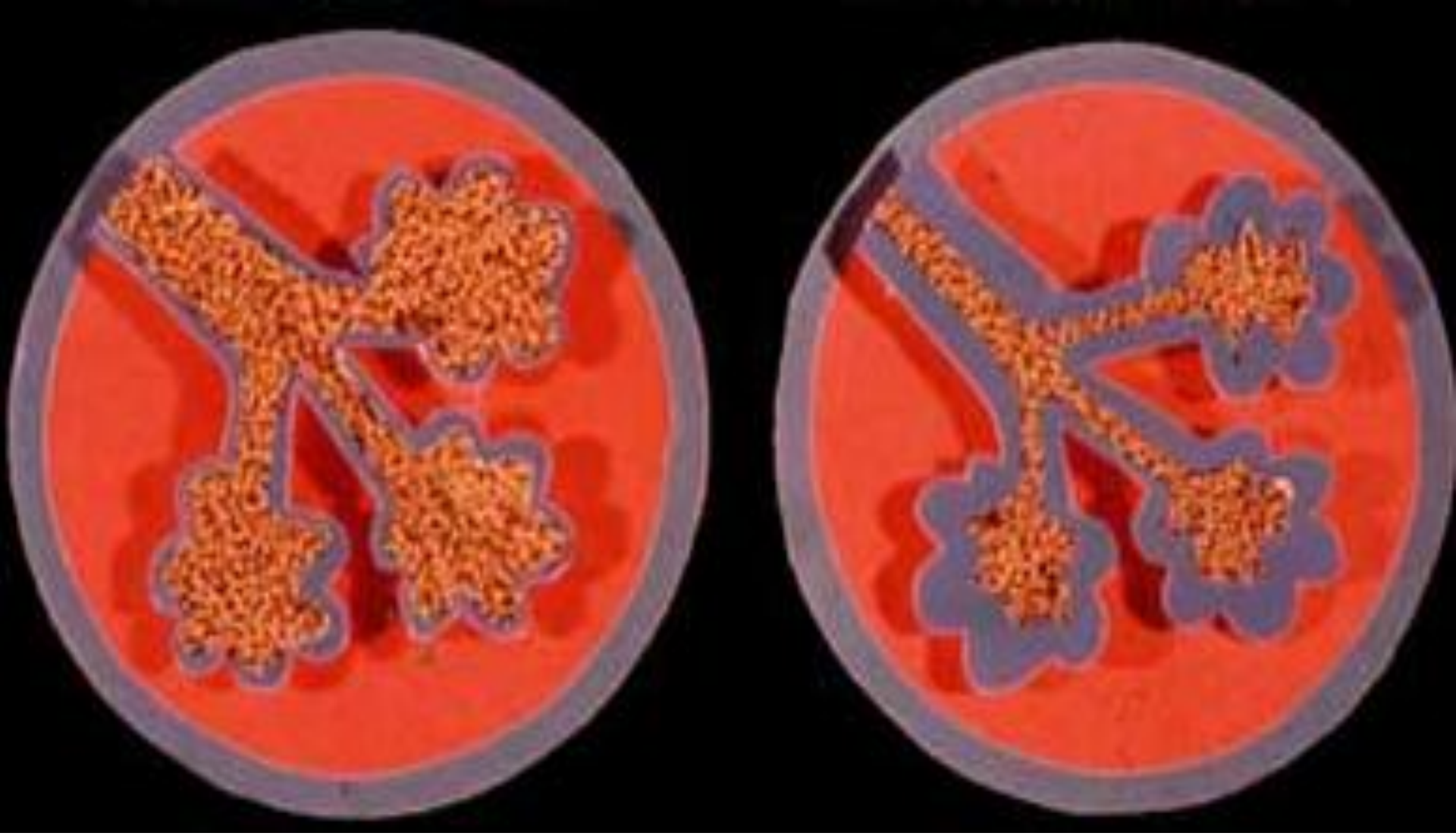
”small sized” lungs



Extensive fibrosis with emphysematous changes and great pleural thickening: visceral, parietal, and diaphragmatic. Lower lobe predominantly involved

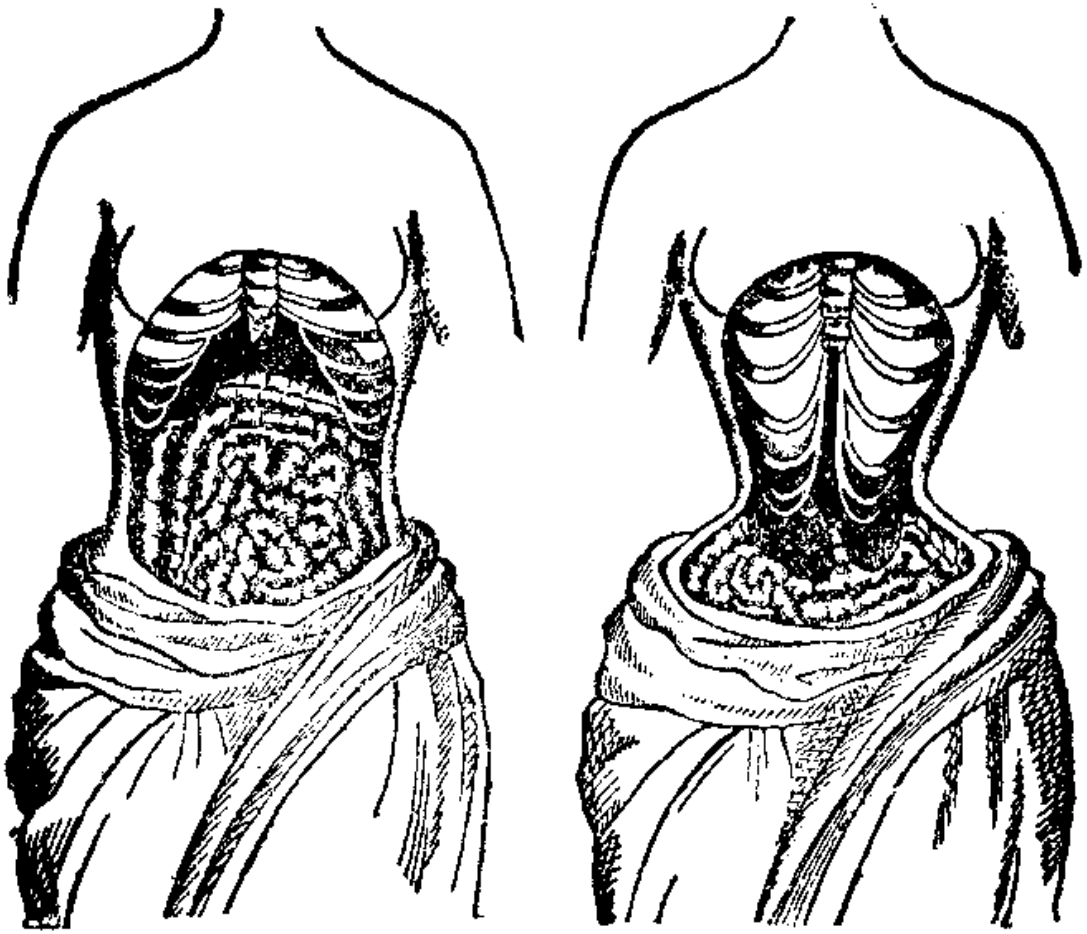

Alveoli
Alveolus
Interstitium
Capillary Bed


Trachea
Bronchus
Bronchiole
Alveoli
Bronchiole
Alveolus
Interstitium
Capillary Bed



The principle of lung fibrosis

Important -Restriktive lung function



A.

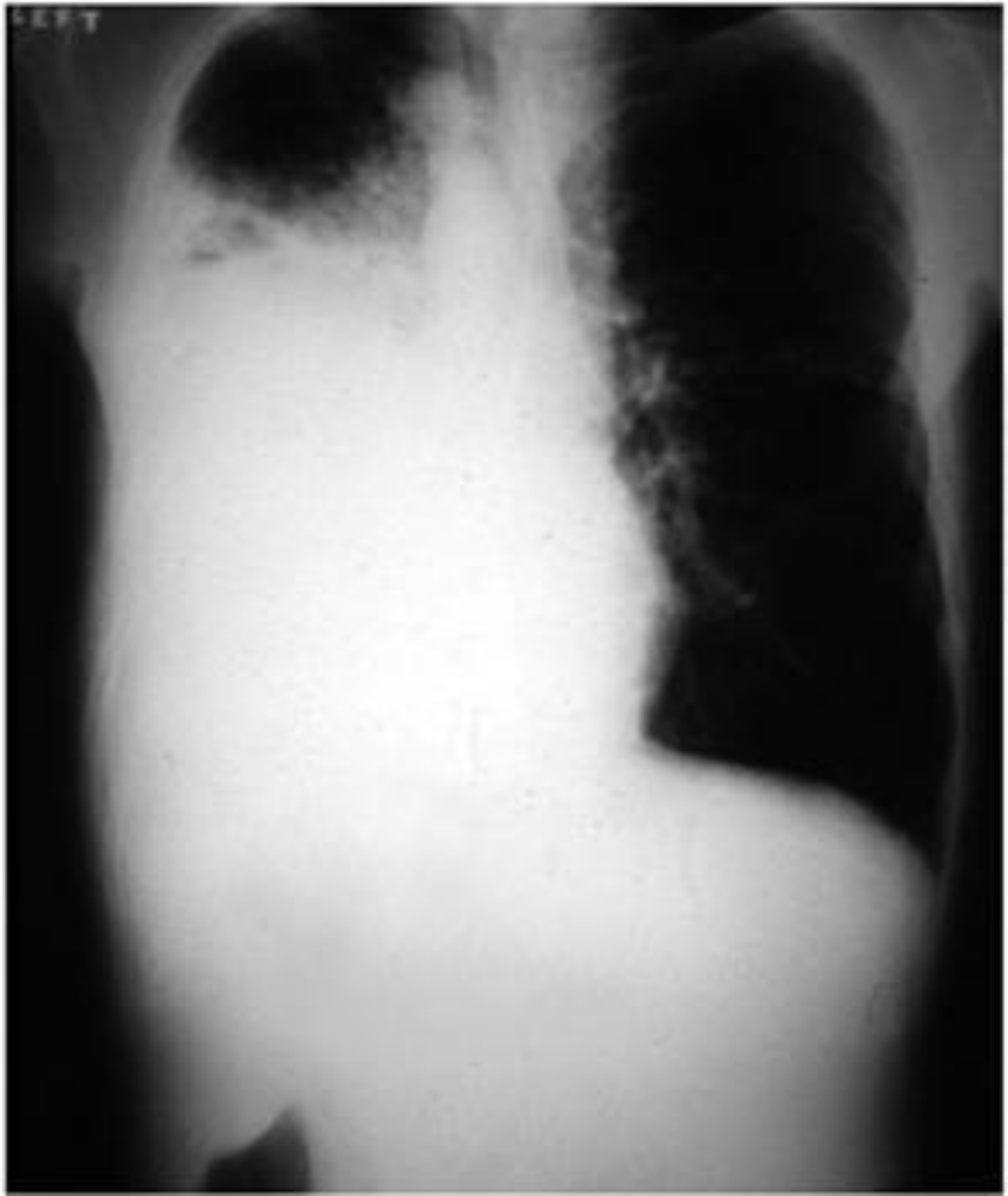
The ribs of large curve; the lungs large and roomy; the liver, stomach and bowels in their normal position; all with abundant room.

B.

The ribs bent almost to angles; the lungs contracted; the liver, stomach and intestines forced down into the pelvis, crowding the womb seriously.

Nature Versus Corsets Illustrated.

Also seen without problems in the lung parenkyma



In principle there is are ”acute” restriktive disorders

Pleuraexsudate, pneumonia
Atelektasis etc..

However. Normally in lungfibrosis

Disease localized in the paremkyma

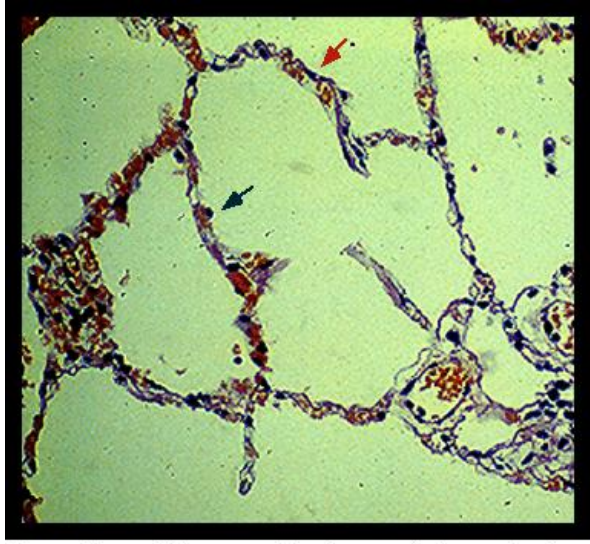
What is lost is forever lost …..

…if treatment is not started in time…

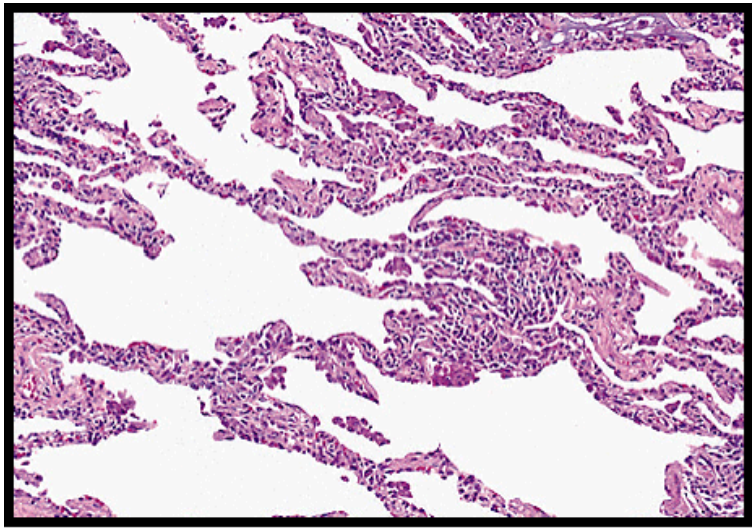
What is ILD or DPLD ?

- Diffuse parenchymal lung diseases, often collectively referred to as the interstitial lung diseases (ILDs), but many names are existing for the same disease leading to a lot of confusion in the area

- The term *interstitial* is misleading since most of these disorders are also associated with extensive alterations of alveolar and airway architecture



Normal lung High power photomicrograph shows alveoli

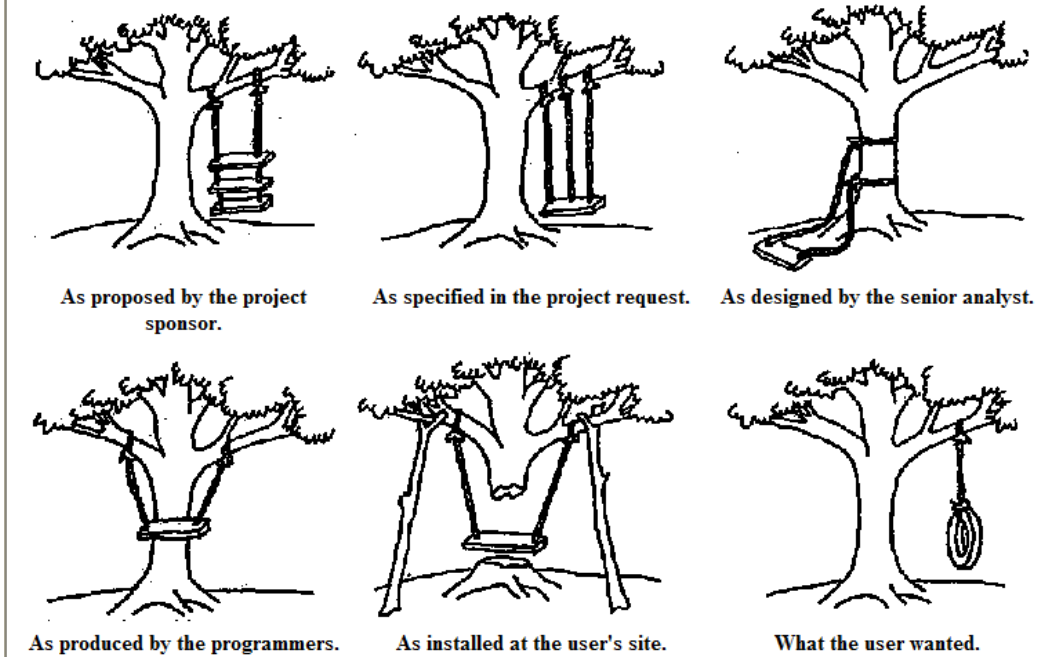


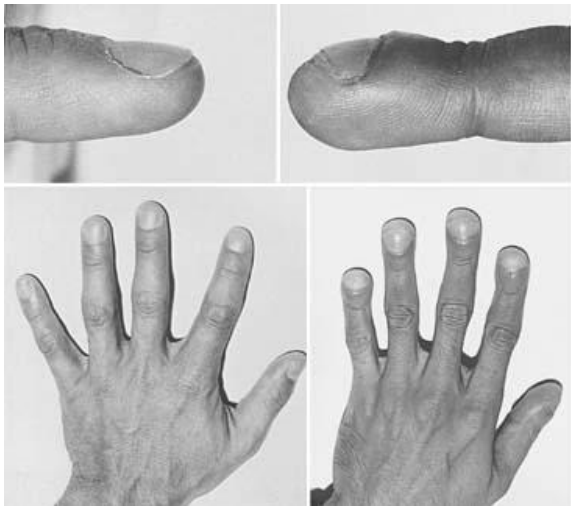
Nonspecific interstitial pneumonia, cellular pattern The

Symptoms

- Signs
- Slowly progession
  - But attracts is prevalent
- Breathlessness
  - At first at activity
  - Later all the time
- cough
  - Non- productive

- Findings cyanosis
  - Low saturation
- low lung function
  - TLC
  - RV
  - dlco
- Dromstikfingers
- Velcro sound at stethoscopy





Findings
Lung Function

-overall

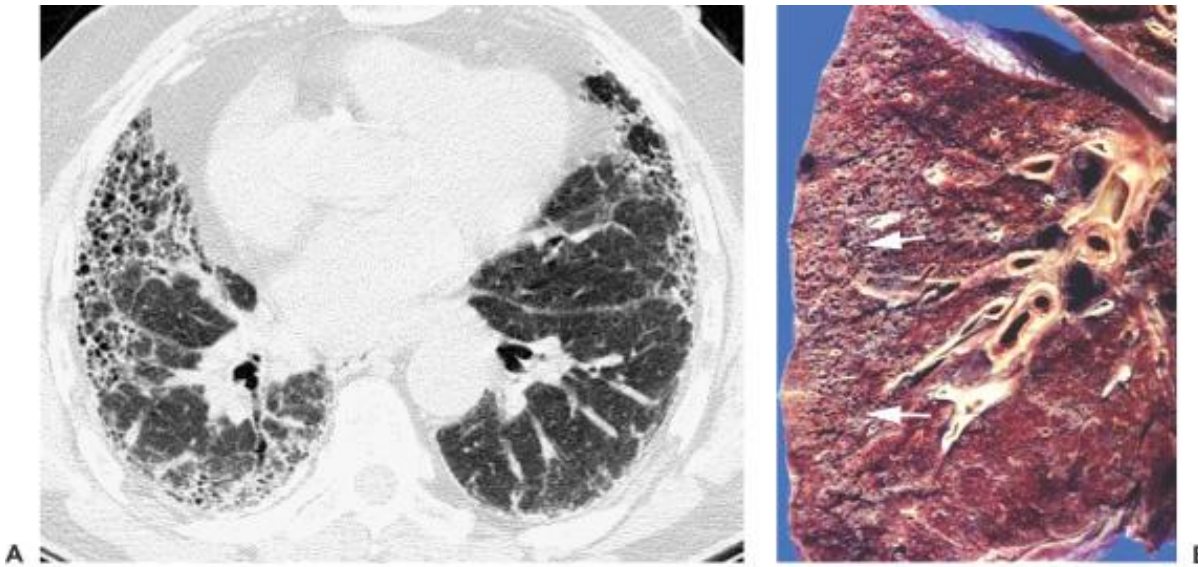
Spirometry reveals a restrictive pattern.

FVC is reduced, but FEV_1/FVC normal/supernormal.

All lung volumes –

TLC, FRC, RV –

Are Reduced.



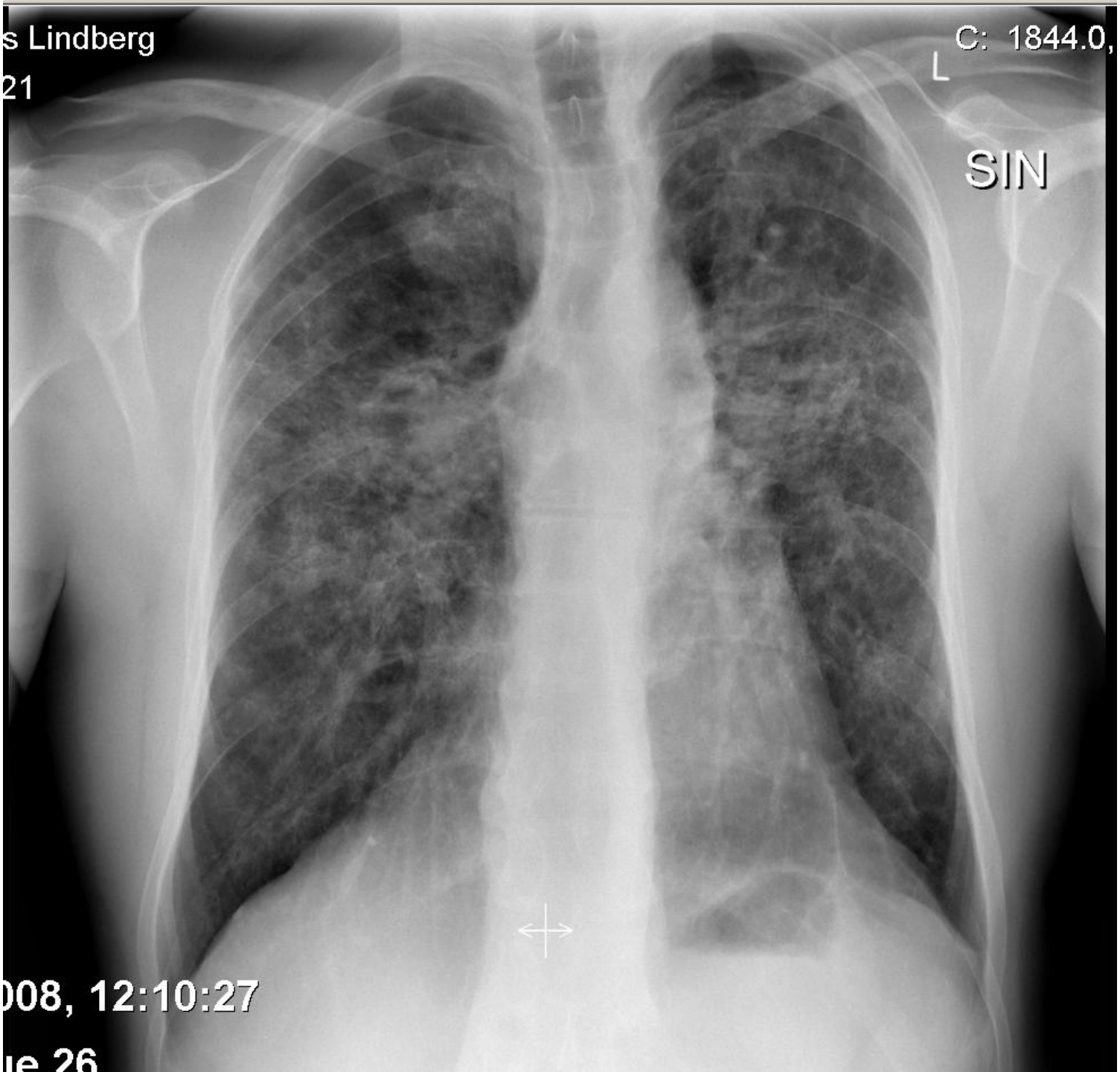
asbestosis

CASE

1975, operatted for retentio testis. .
1989, operated inqvinal hernia right side. .

47 year old male: The Pt. s symptoms started 7 month ago with dry cough, initially treated by a doctor with Salbuvent mixture, but as no effect . Never smoker.
He then stopped the treatment after 3 days due to heart beat and tremor. Cough is mostly dry but intermittent yellow. Had has som pain in the joints without swelling.

The patient has a complain of increasing dyspnea especially the last month. Whezzing has been observed a few times. Some joint pain especially in both shoulders.
Returns to his doctor after 7 month and the doctor due a X-RAY: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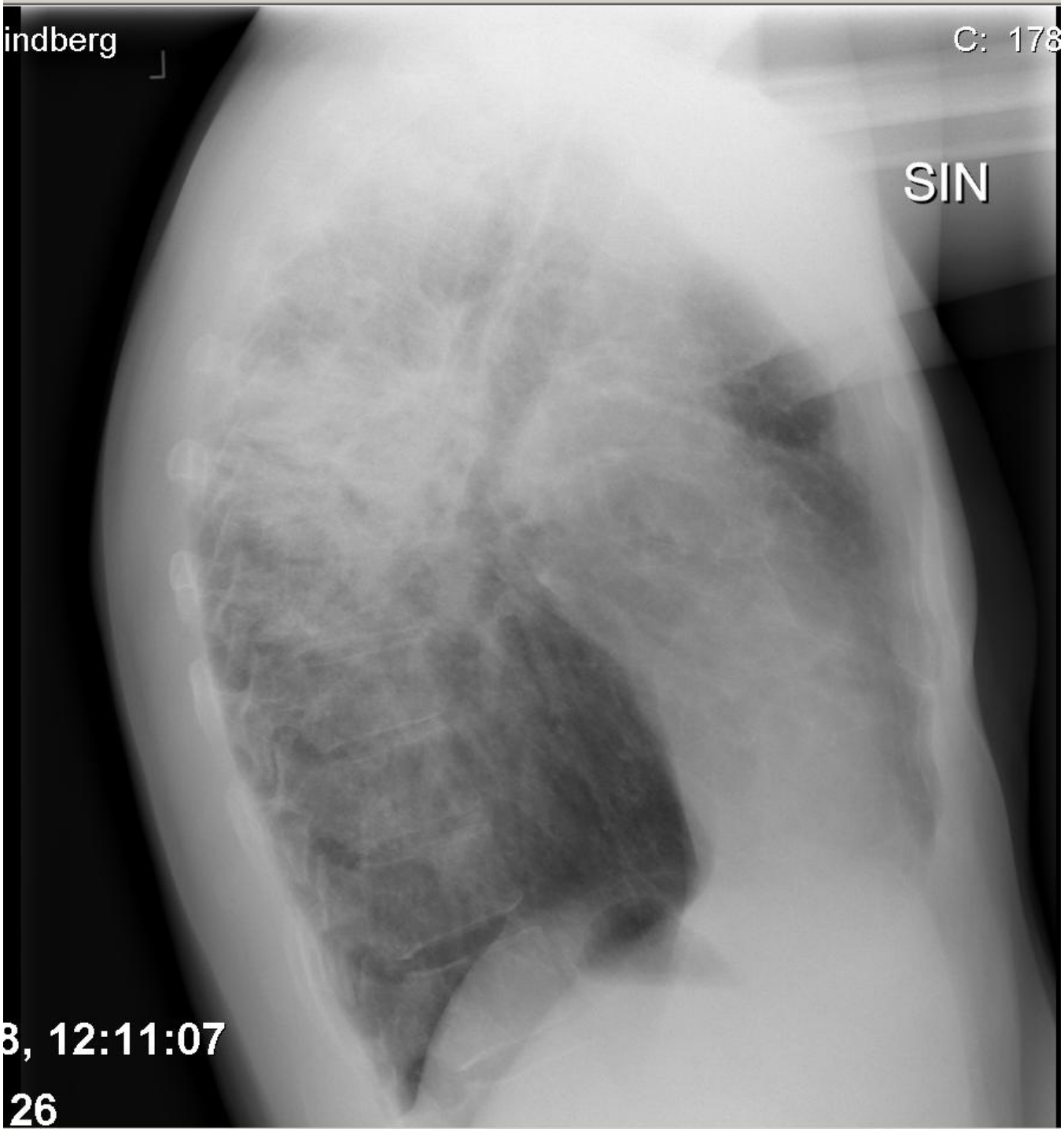
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26

CASE

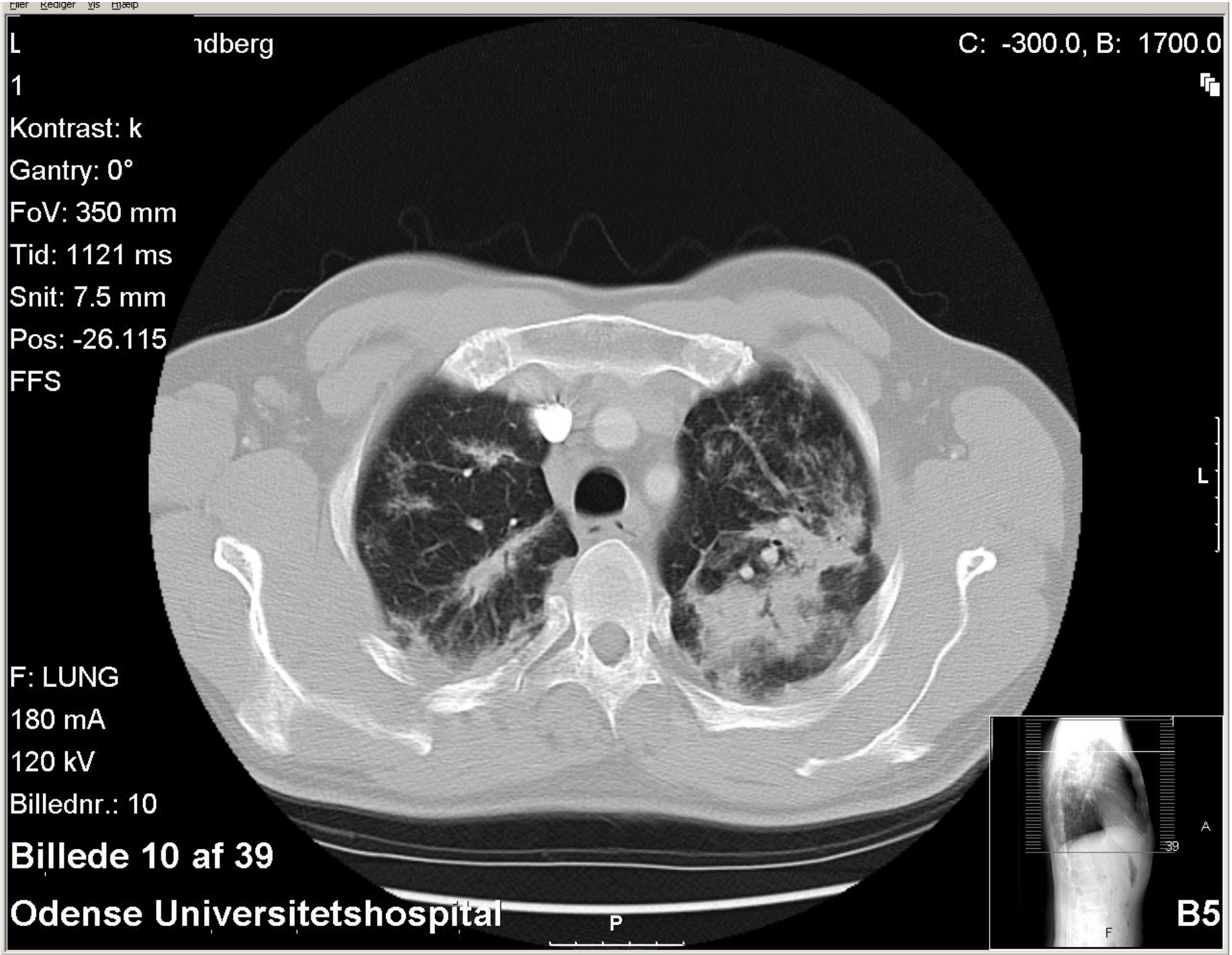
X-RAY showing **bilat. Infiltrats and increased size of lympnodes at hilus.**

The doctor says pneumonia and gives Klacid 500mg x2

Do you agree ??

How do we approach this patient ?

- Tentative diagnoses
  - Asthma
  - Infection
  - Diffus parenkymatøs lung disease
    - In particular sarcoidosis
  - Cancer
    - lymphoma
- Tests
  - CT- thorax + upper abdomen
  - Bloodtest
  - Lung function incl reversibility
  - Bronkoskopi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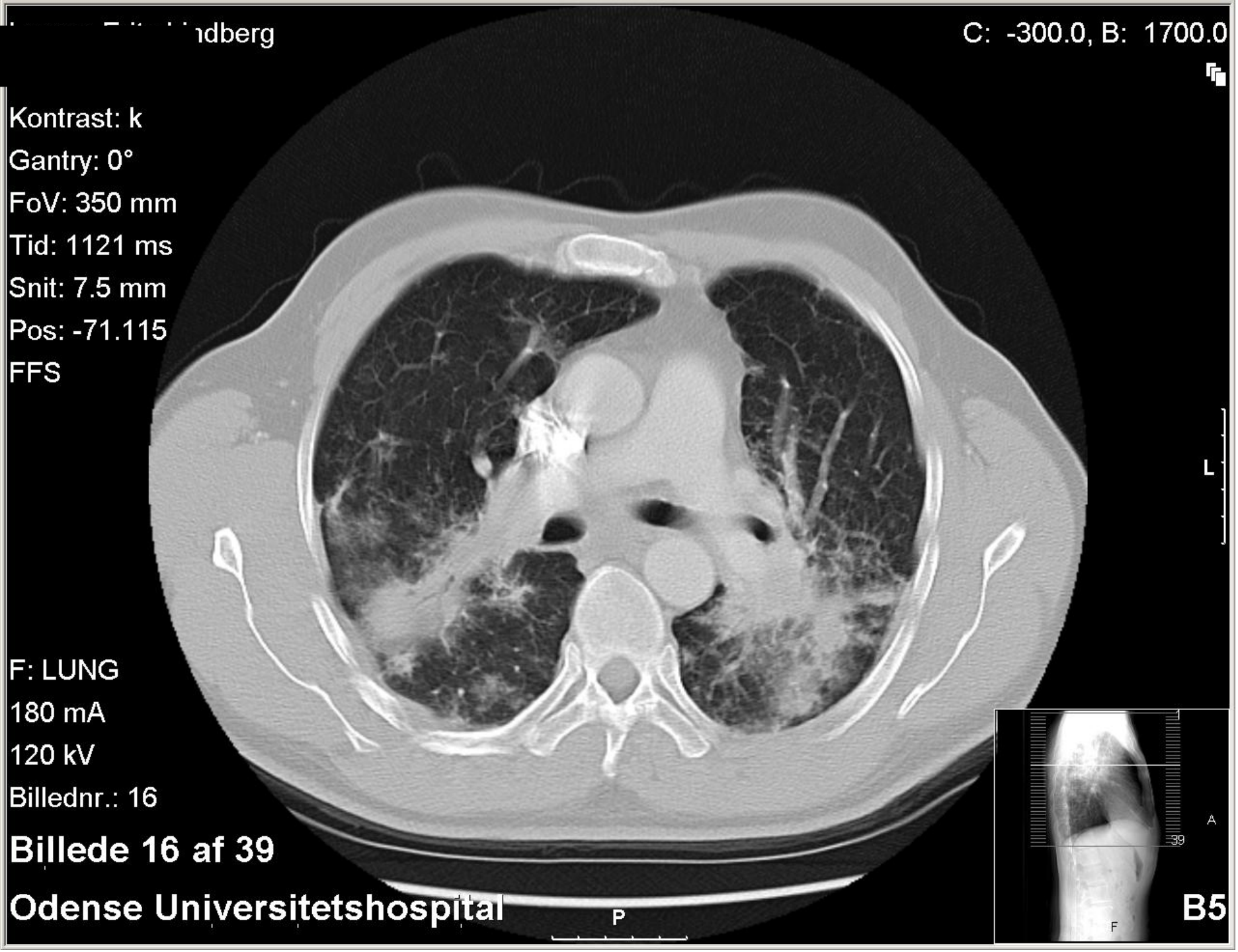
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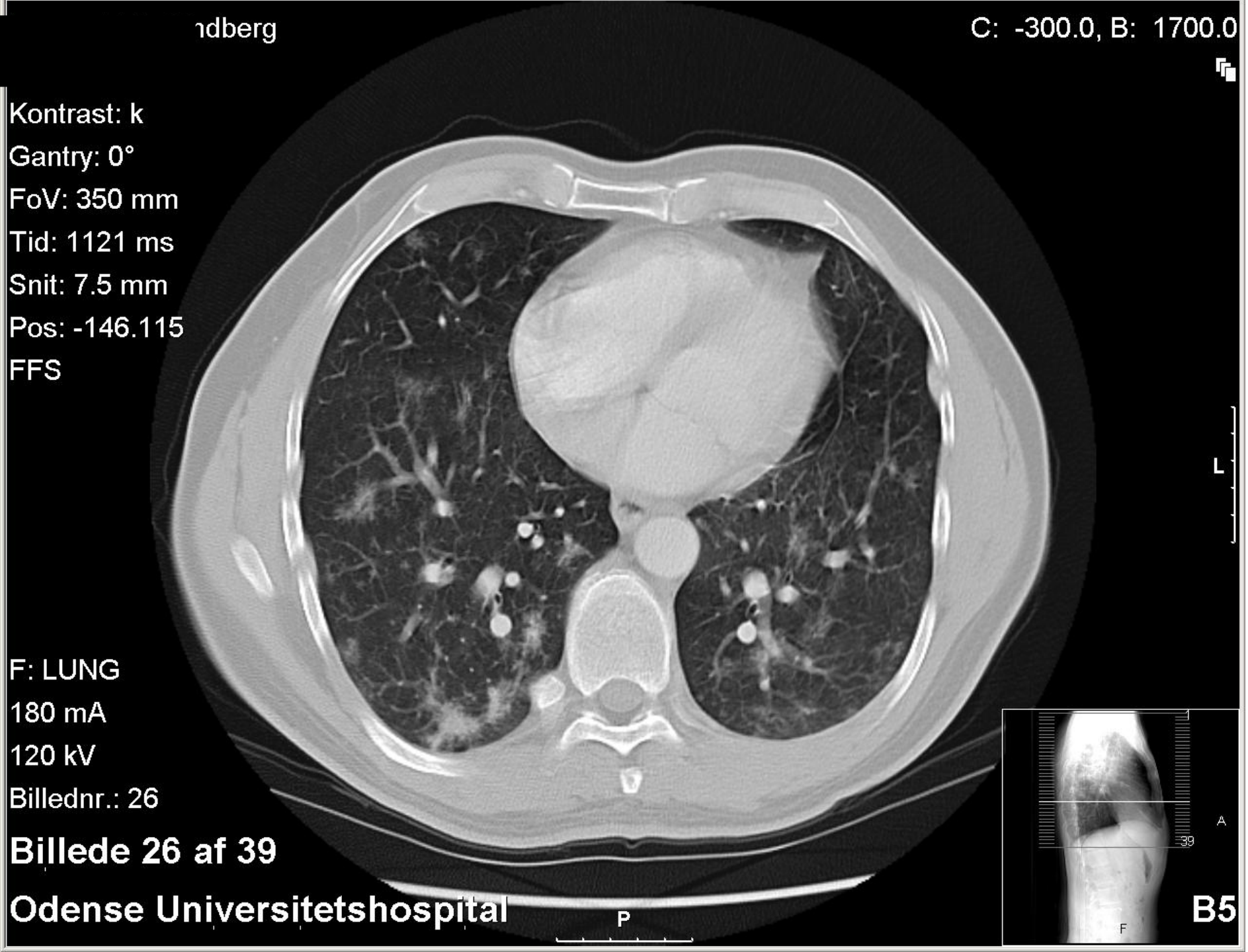
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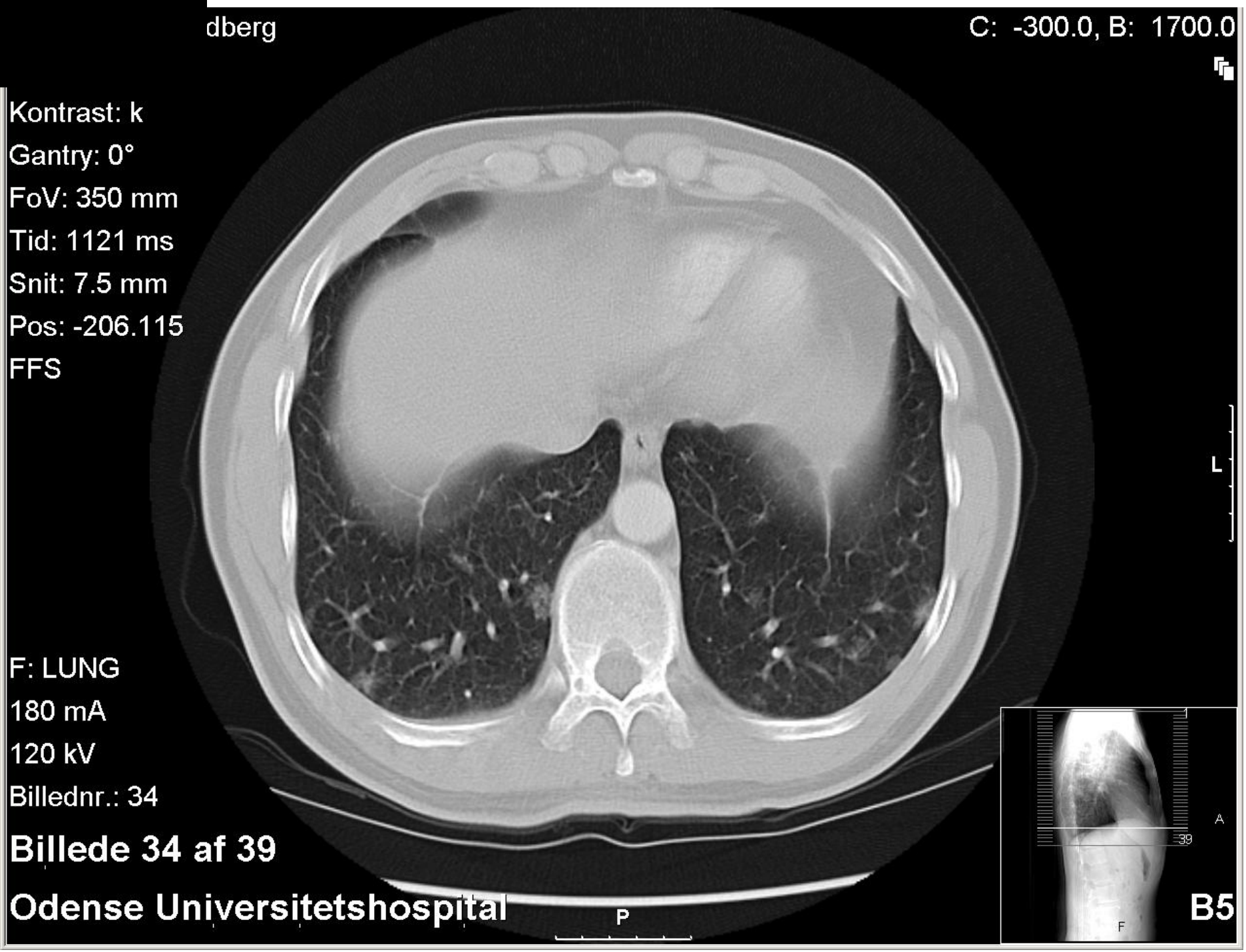
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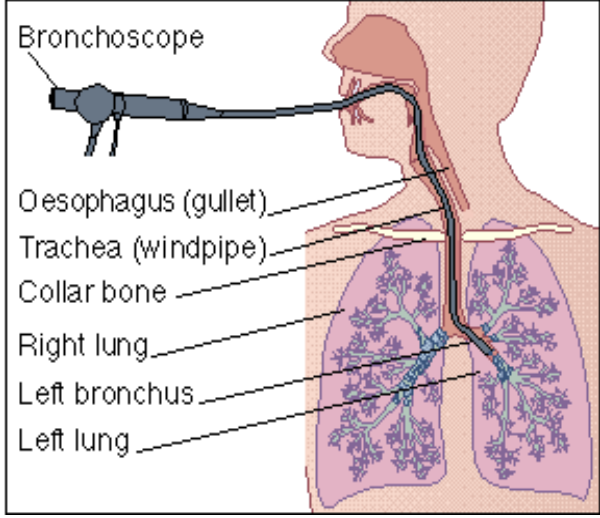
Analysenavn	Resultat	r	C	R	Enhed	Ref.lav	Ref.høj	T	Kode	P
Hæmoglobin;B	9,1	☐			mmol/l	8,0	11,0		HB	6
Erytrocytmorf	~~~~~~~~~~	☐							ERYMRF	6
Leukocytter;B	9,1	☐			10E9/l	3,0	10,0		LKC	6
Leukocyttype		☐							LEUTY	6
Neutrofile	7,83	☐		*	10E9/l	1,50	7,50		NEUTRO	6
Eosinofilocytter	0,18	☐			10E9/l	0,04	0,50		EOSINO	6
Basofilocytter	0,00	☐			10E9/l		0,20		BASO	6
Lymfocytter	0,55	☐		*	10E9/l	1,00	3,50		LYMFO	6
Monocytter	0,55	☐			10E9/l	0,20	0,80		MONO	6
Leukocyttype kom	~~~~~~~~~~	☐							LCTKOM	6
Kreatinin;P	94	☐			µmol/l	62	134		CREA	6
Calcium-ion;S	1,29	☐			mmol/l	1,19	1,29		CAI	6
LD	168	☐			U/l	105	205		LD	6
ALAT	21	☐			U/l	10	70		ALAT	6
BASP	43	☐			U/l	35	105		BASP	6
CRP	<5	☐			mg/l		10		CRP	6
ACE	!KOMM	☐	C		U/l	30	115		ACE	6

ACE= 94 og ANA,ANCA;Anti-CCP: normal; IgG,IgM,IgA,IgE normal
EKG= SINUS RYTME no ischemia

LF is technically ok performed; FEV1 =3,14 (65% of predicted), FVC= 4,51 (73% of predicted)
A ratio of 70 ?

Bronkoscopy was done



clinic:

cough

Xray; diffus infiltrations, also seen on CT.

Bronkoskopi: no tumores, mucosa east to bleed.

A: biopsy (Transbronkial) right upper lope

MAKROSKOPI

A: many light very small we try to make some out of it, one tissue done 1 kps/grh

MIKROSKOPI BESKRIVELSE:
Bronkiebiopsiy, surfase has normal respiratorisk
epitel without atypi. Under the epitel mange epiteloidcel-
Granulomea with giantcells. No amyloid. No nekroting granulomatous inflammation,
Why consider diseases with that area . No sign of malignency.

/dpa

-

Gunvor Madsen

Culture: D+R normal flora
PCR was all negative

TEST: Among the diseases mentioned below where do you see Non-caceating granulomas inflammation ???

1. COPD
2. Asthma
3. Idiopatic lungefibrosis
4. Sarcoidose
5. Allergic alveolitis
6. Lymfoma
7. Tuberculose
8. Wegeners granulomatosis

Among the diseases mentioned below where do NOT see non-caceating granulomas inflammation ??? (casuistik not included)

4, 5, 6 og 8 are the right one

1. COPD
2. Asthma
3. Idiopatic lung fibrosis
4. Sarcoidose
5. Allergic alveolitis
6. Lymfoma
7. Tuberculosis
8. Wegeners granulomatosis

More tests for our patients ?

TLC, RV; DLCO;
6 min walking test

30.04.04 we found:
TLC: 87 % of predicted
RV: 140 % of predicted
Diffusionscapacity is decreased to 63 % of predicted
FEV 1: 63 % of predicted.
FVC 73 % of predicted.
The Pattern is slight obstruktive as FEV1/FVC ratio is 70%.

Pt's 6 min. Walking test shows saturations between 93% - 96% and distance of 470 meters. 1 - 2 on Borgs dyspnea skale before test and 4 – 5 after the 6 min..

Diagnosis ?

Resume: 47yrs male cough, dyspnea, joint pains, XRAY shows promint hili, with increased lymfenodes and infiltrative changes in the parymkyma of the lung primarily in hilus and apicalt, FVC 70% of predicted, Peak flow variability is normal. non-caceating granulomas inflammation

Diagnosis ?

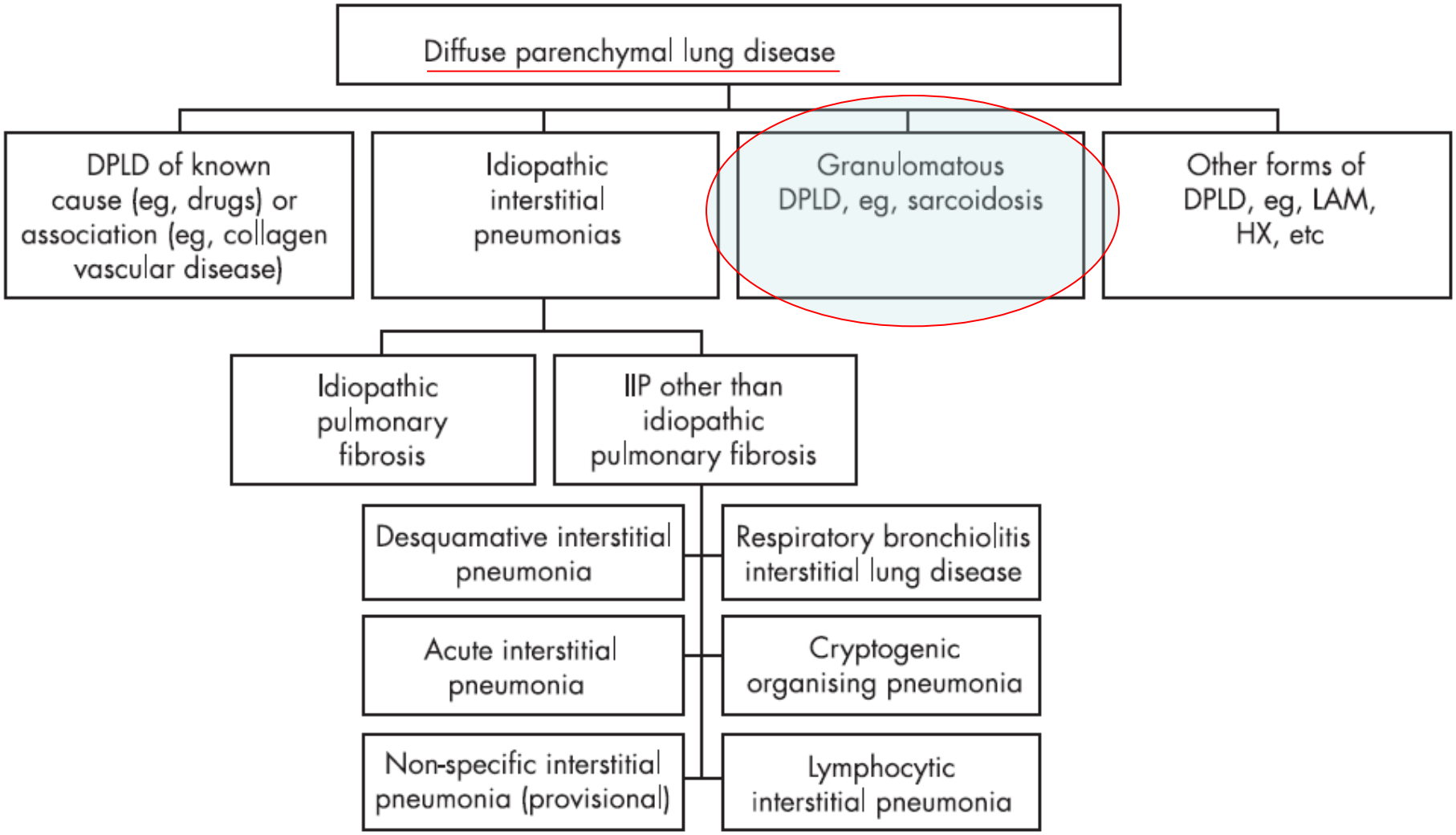
1. We need further test as we can not be sure yet
2. The Patient has sarcoidose
3. The Patient has lymfoma
4. The Patient has asthma
5. The Patient has histeocytosis X
6. The Patients allergic Alveolitis
7. Wegeners granulomatose

Resume: ~40-årig mand hoste, dyspnø, ledsmerter med prominerede hili, forstørrede lymfeknuder samt infiltrative forandringer hilært og apicalt i lungeparenkymet, Peakflow variabilitet ~10%;

The patient disease : 2 is right

1. We need further test as we can not be sure yet
2. The Patient has sarcoidose
3. The Patient has lymfoma
4. The Patient has asthma
5. The Patient has histeocytosis X
6. The Patients allergic Alveolitis
7. Wegeners granulomatose

Diseases


Diffuse parenchymal lung disease
DPLD of known cause (eg, drugs) or association (eg, collagen vascular disease)
Idiopathic interstitial pneumonias
Granulomatous DPLD, eg, sarcoidosis
Other forms of DPLD, eg, LAM, HX, etc
Idiopathic pulmonary fibrosis
IIP other than idiopathic pulmonary fibrosis
Desquamative interstitial pneumonia
Respiratory bronchiolitis interstitial lung disease
Acute interstitial pneumonia
Cryptogenic organising pneumonia
Non-specific interstitial pneumonia (provisional)
Lymphocytic interstitial pneumonia

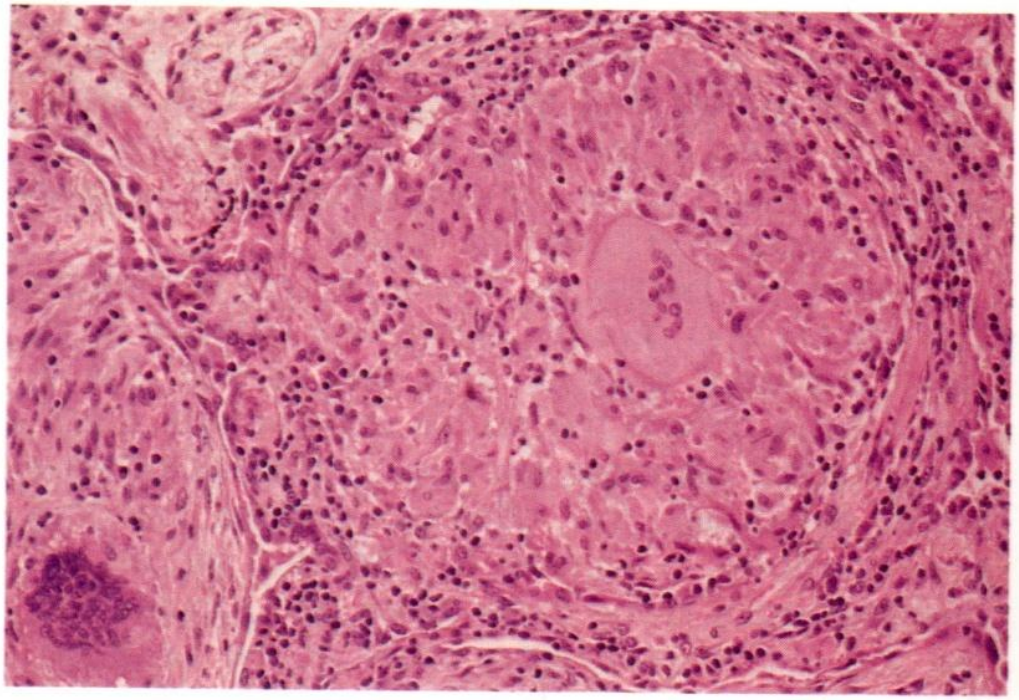
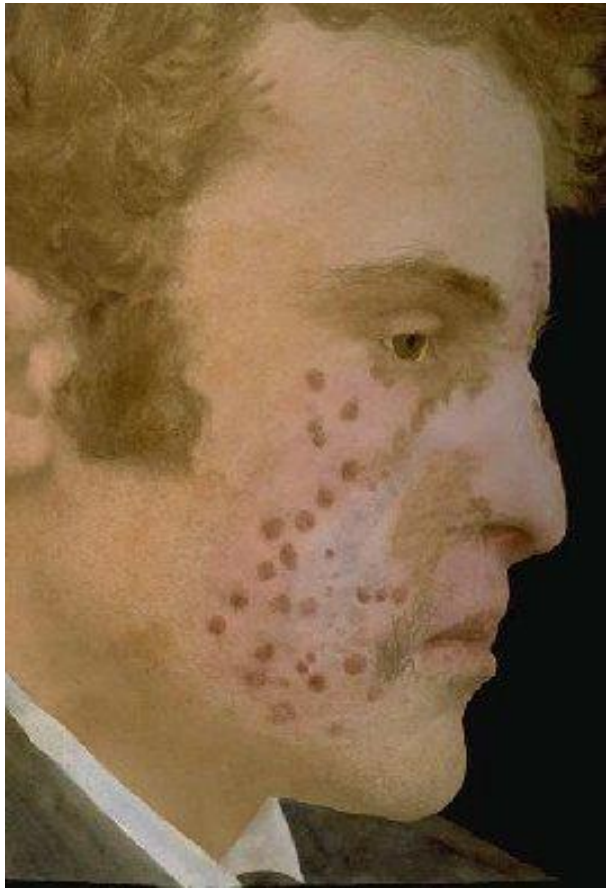
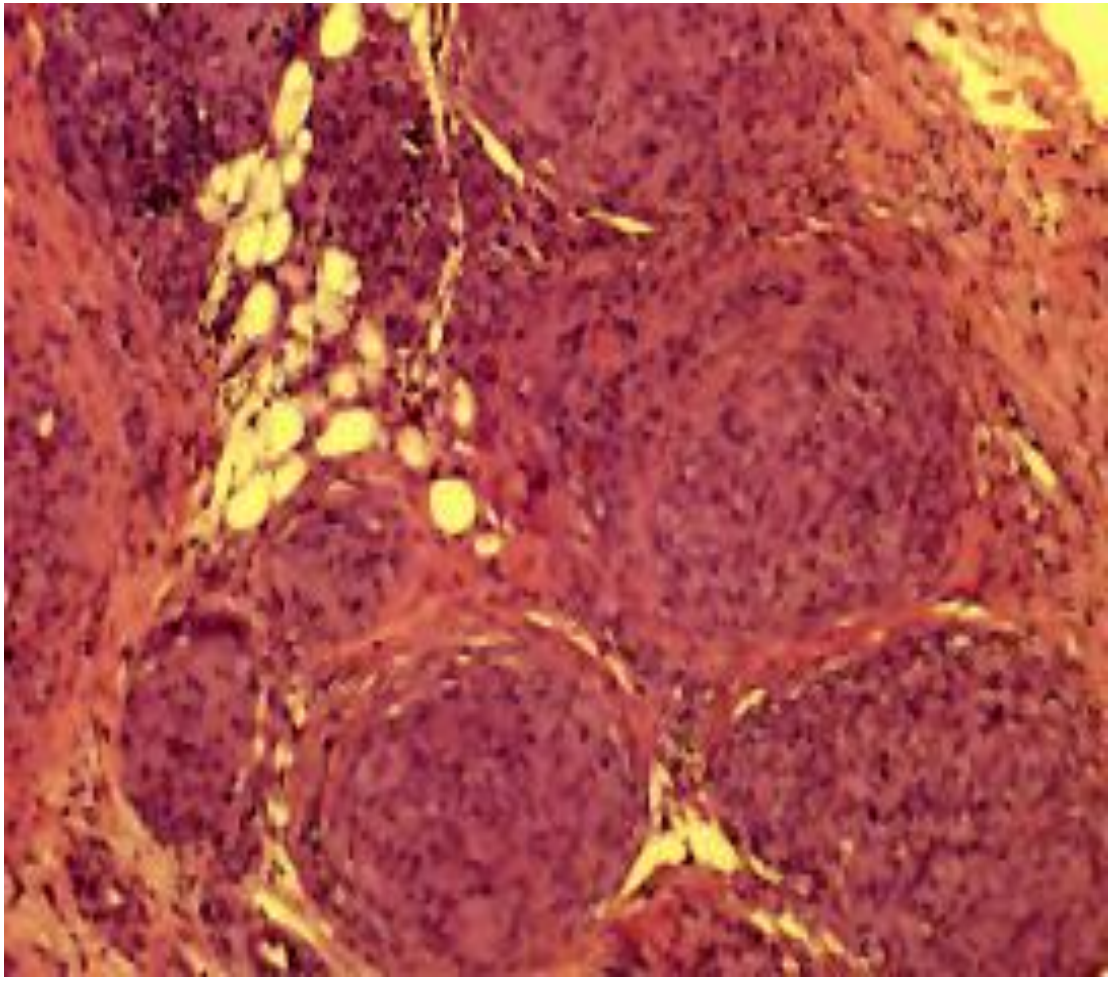


Figure 15–33. Characteristic sarcoid noncaseating granulomas in lung with many giant cells. (Courtesy of Dr. Ramon Blanco, Dept. of Pathology, Brigham and Women's Hospital, Boston.)



Sarcoidose



Patologi:
Non-caceating granulomas disease

Who gets Sarcoidose??

Most often between 25-40 yrs

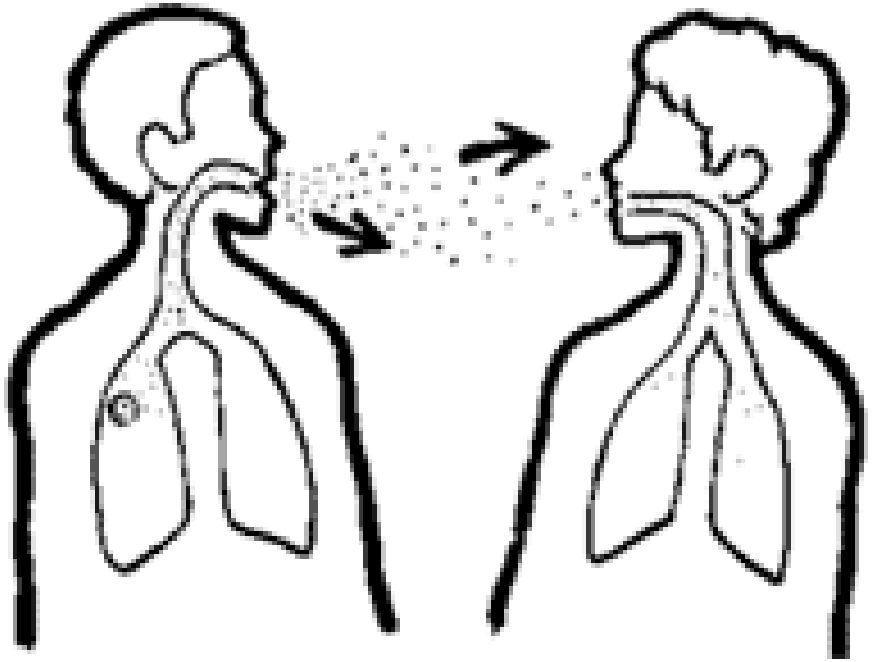
More women than men

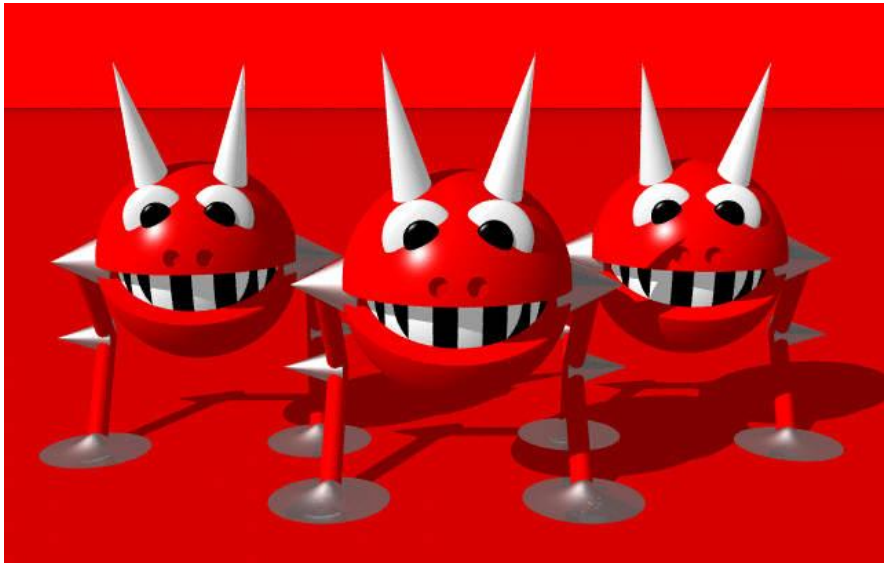
More negro than white

Is Sarcoidose infectious?

NO !

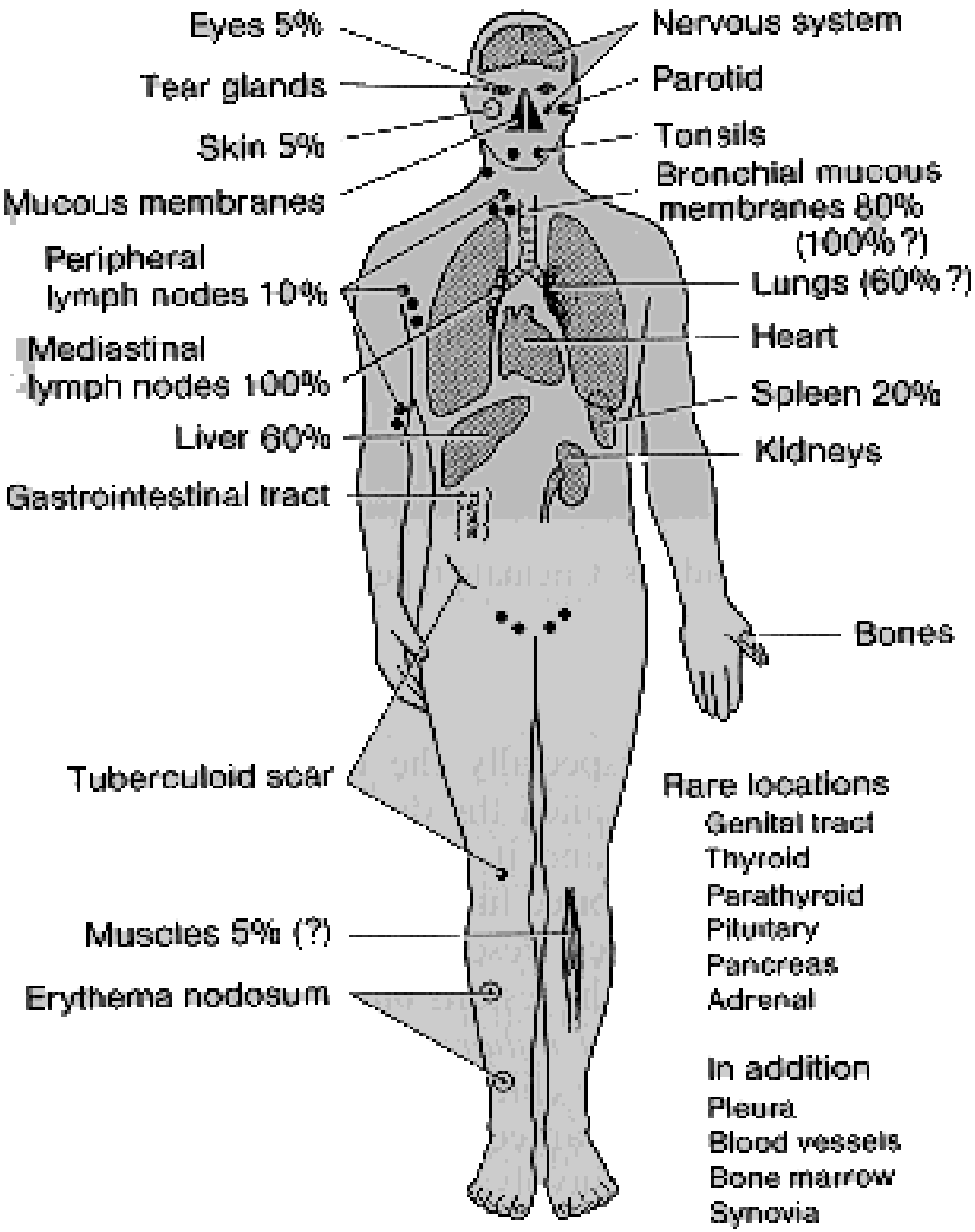
Familiar cases seen !





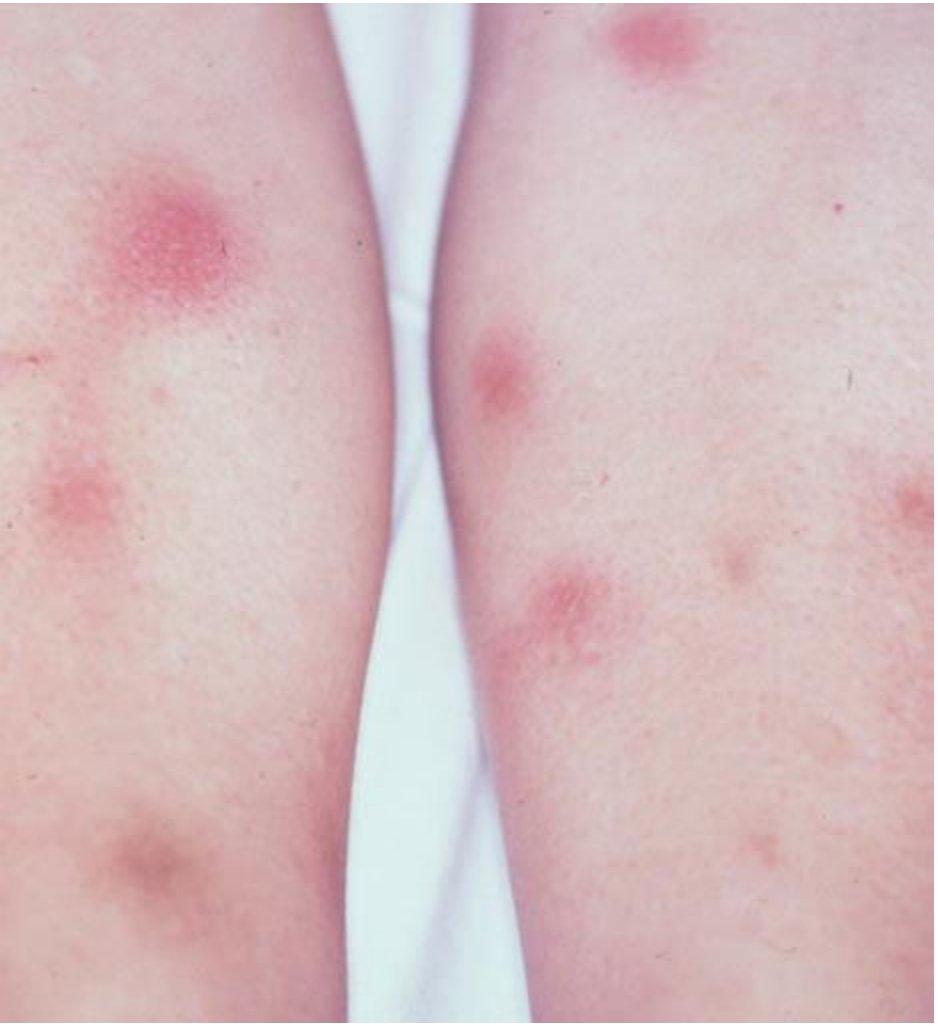
Klinik

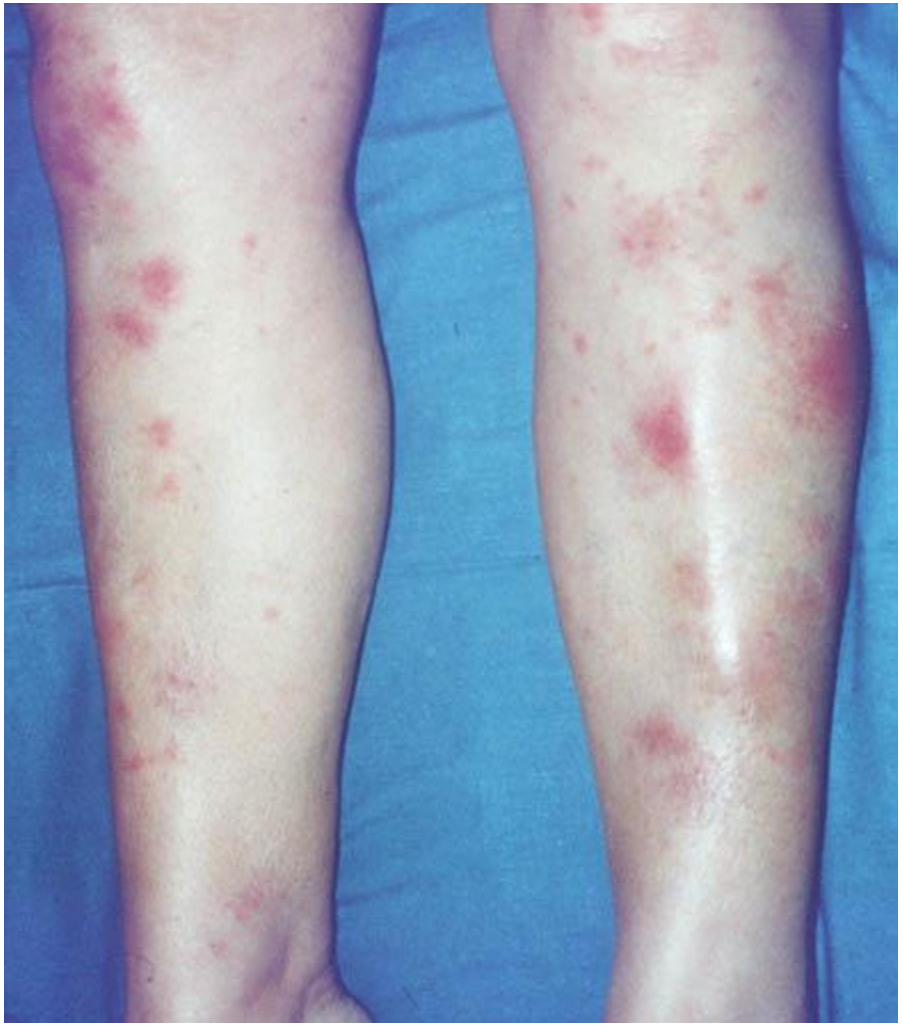
Multiorgan disease



Lung symptoms
>90%

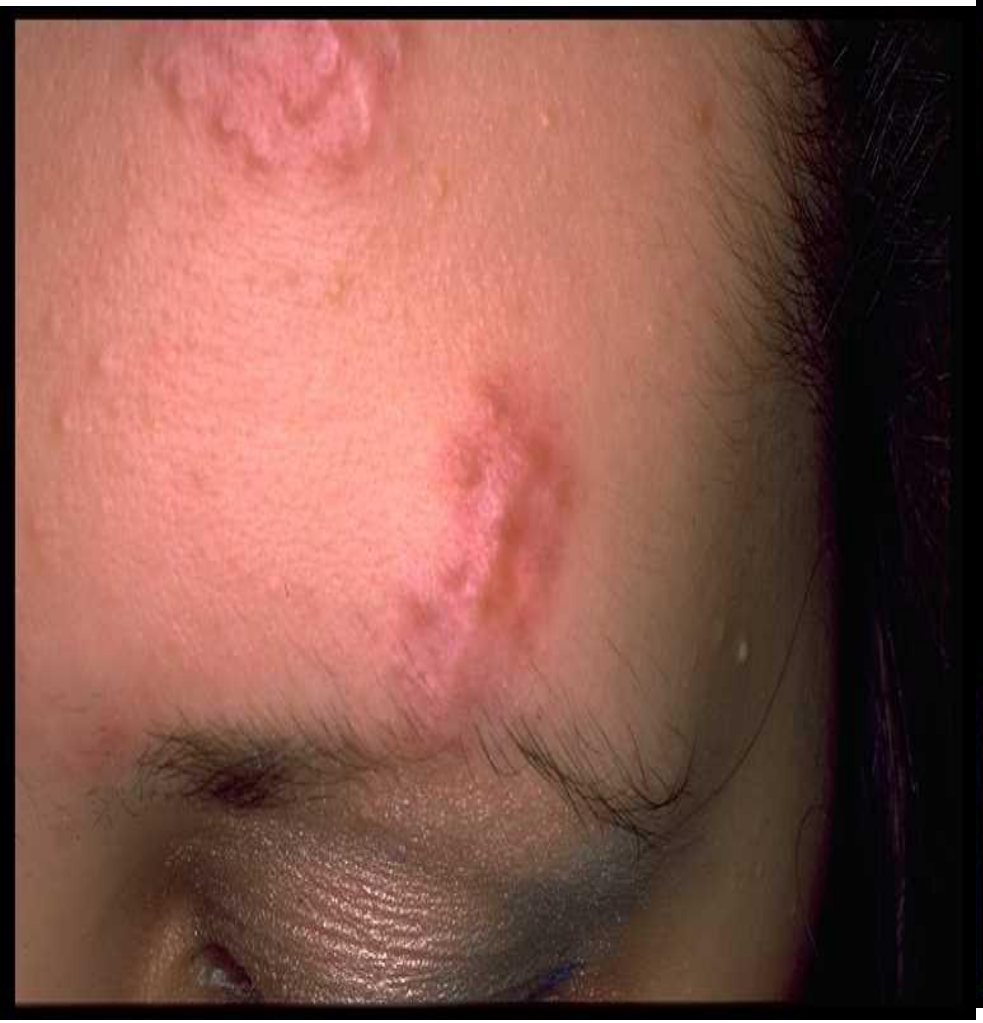
Cough
Dyspnoe
Chest pain

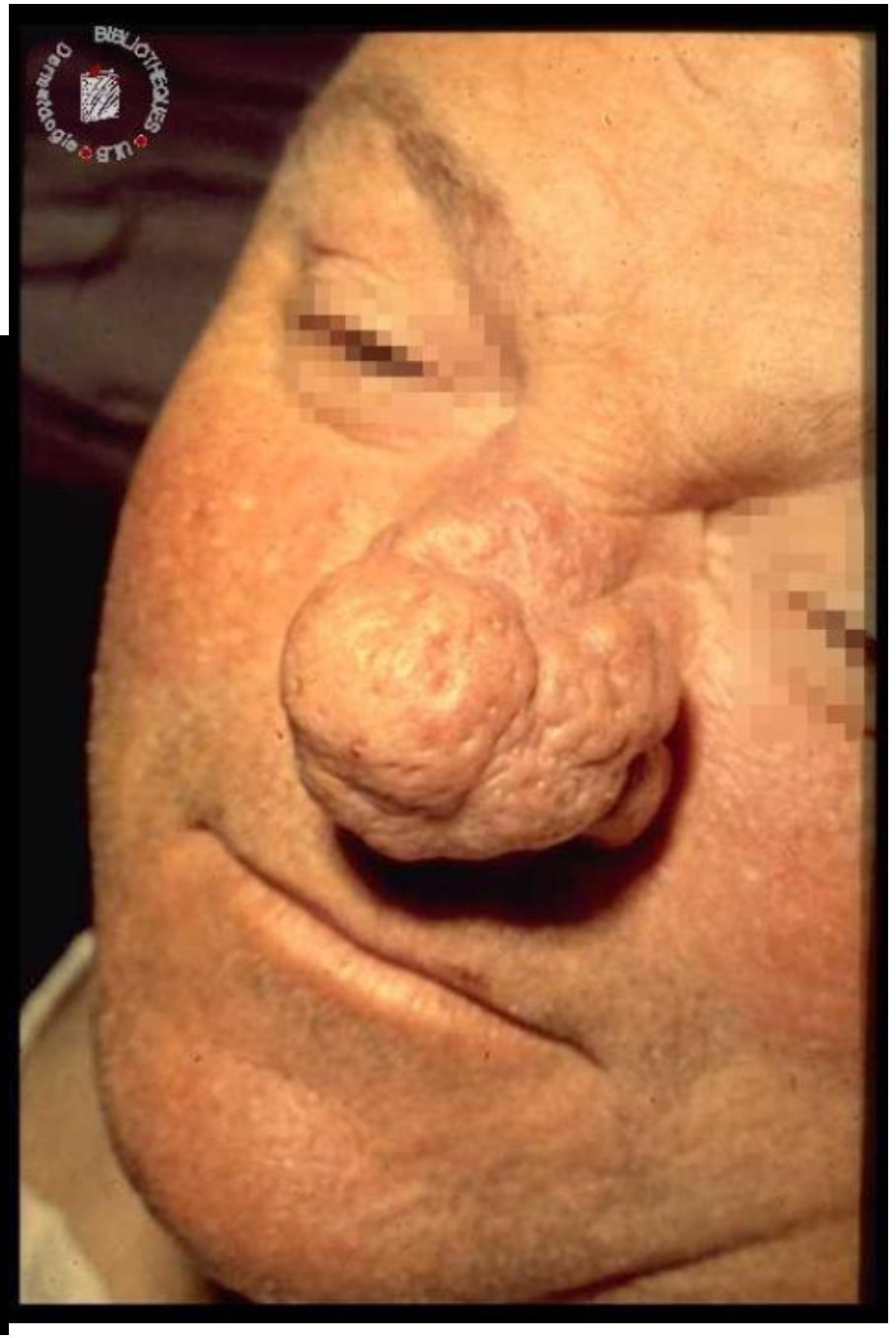




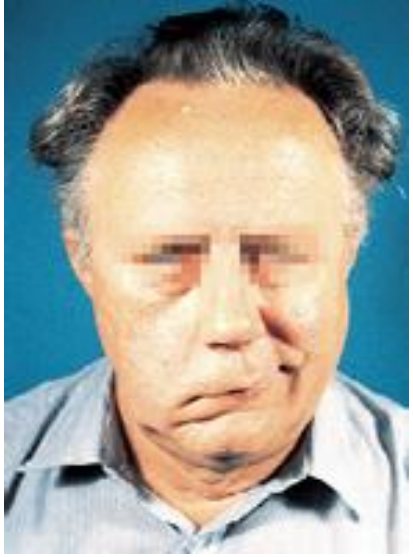
Skin changes: Erytema nodosum

Skin Changes

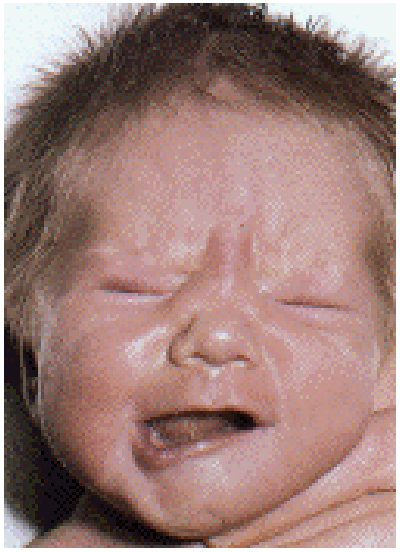




CNS manifestations

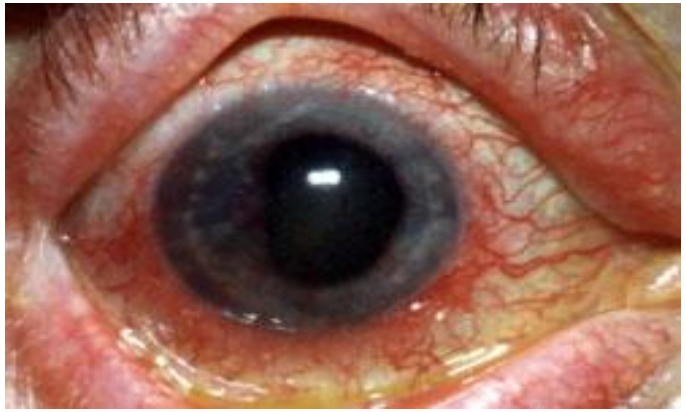


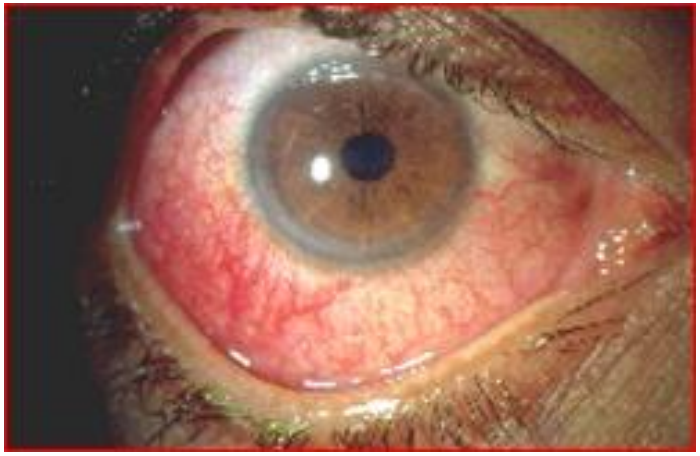
10% a debut of parese
Often perifer Fasialis parese

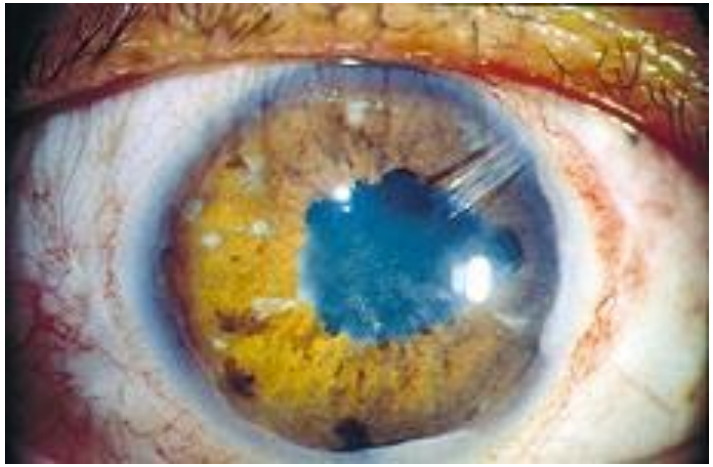


Eye symptoms

Send to eye doctor







Sarcoidose treatment

Steroids

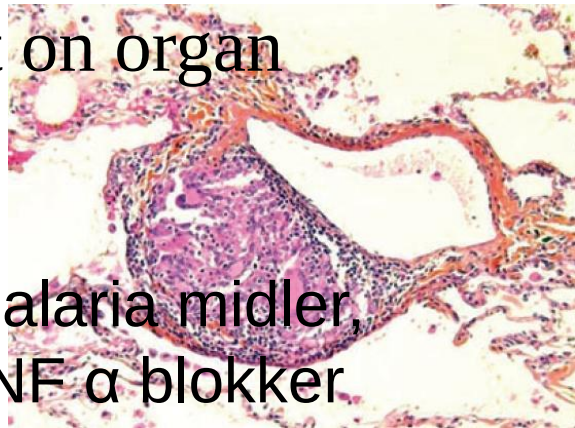
Effect on symptoms +++++

Effect on lung function +++

Effect on prognose +?

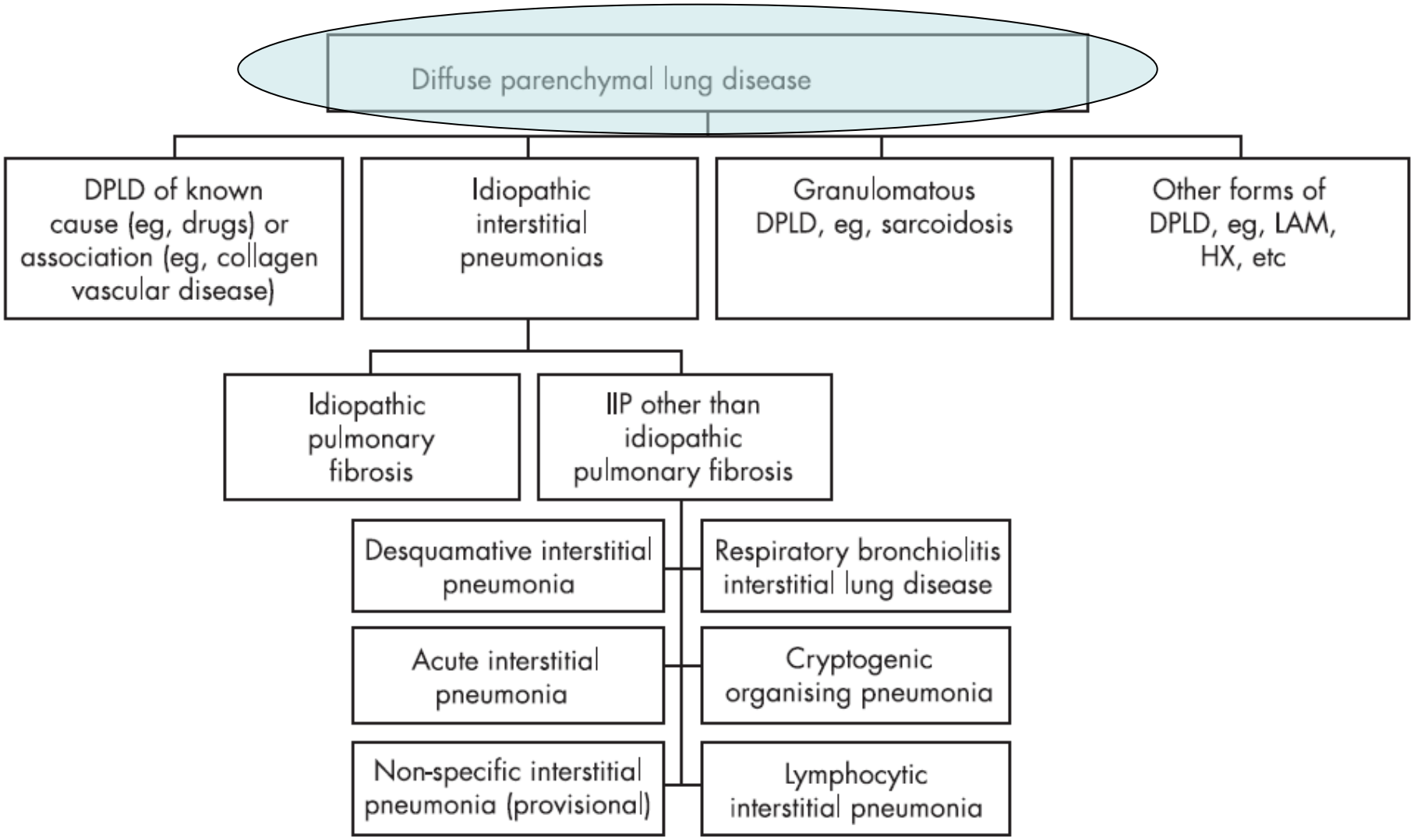
Steroid

- <u>Systemic steroids</u>
- Lokal – lokal steroid dependent on organ
- Hypercalciema

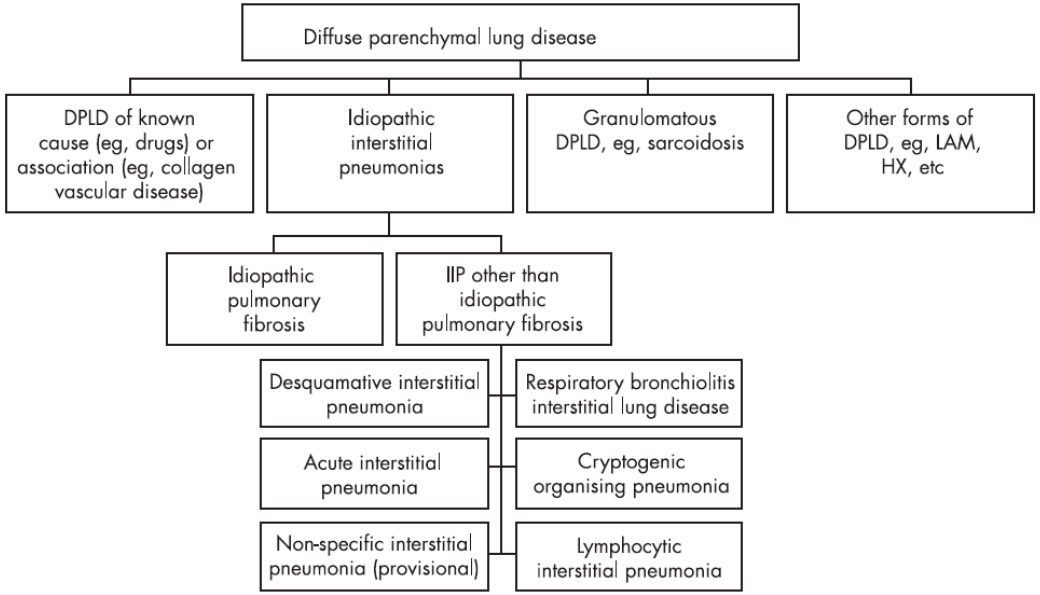


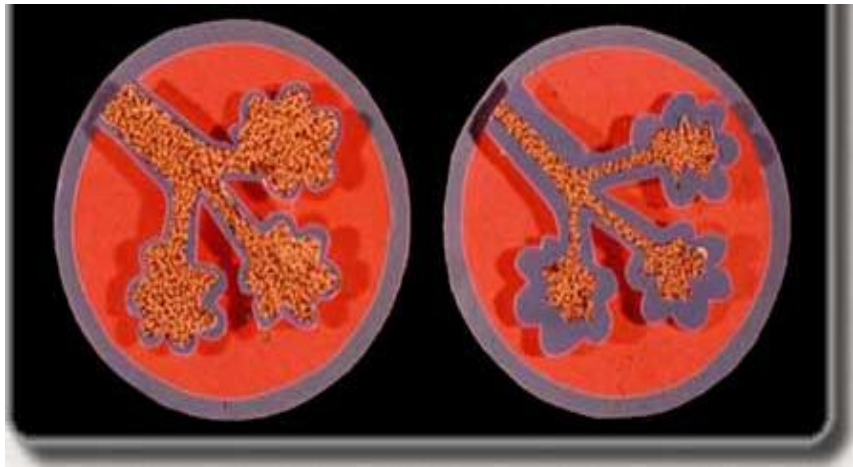
More rare treatments:

Andre Immun modulerende stoffer :anti-malaria midler, Metrotrexat, Azathiprine, Infliximab – TNF α blokker


Diffuse parenchymal lung disease
DPLD of known cause (eg, drugs) or association (eg, collagen vascular disease)
Idiopathic interstitial pneumonias
Granulomatous DPLD, eg, sarcoidosis
Other forms of DPLD, eg, LAM, HX, etc
Idiopathic pulmonary fibrosis
IIP other than idiopathic pulmonary fibrosis
Desquamative interstitial pneumonia
Respiratory bronchiolitis interstitial lung disease
Acute interstitial pneumonia
Cryptogenic organising pneumonia
Non-specific interstitial pneumonia (provisional)
Lymphocytic interstitial pneumonia

- Interstitial lung diseases includes different heterogeous diseases of both known and unknown causes but all with inflammation of the interstitium
- The type of inflammatory respons are different and the degree of progression to interstitial fibrosis



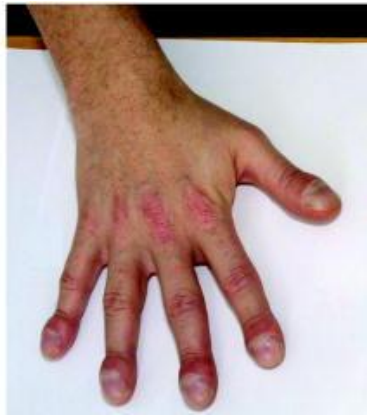


Case

60 yrs old man .The Pt symptoms started for 1,5-2 yrs ago, where he for the first time experienced to get severe dyspnea while running. Dry cough . Still working as leader in and institution for disabled. While work works at least 10 km a day. Ex-smoker, for 20 yrs 20 cigarets a day stopped 15 yrs ago.

Bloodtests showed increased IgG, marginally increased ALT, normal IgM reumafaktor. Normal ACE, anti-CCP, ANCA ,but weakly positive ANA.

Obj: clubbing,, vencro sound on stetocopy



Figur 1. Udpræget clubbing med urglasnegle og hyperæmiske randzoner på højre hånd.

L

6 MINUTTERS GANGTEST

6760 Ribe

Dato 7/12-07 Henvisende læge HDM

Ilttilskud 0 Ganghjælpemiddel -/-

TID	DISTANCE	SATURATION
0 min.	0 m.	91 %
0,37	50 m.	87 %
1,14	100 m.	85 %
1,53	150 m.	81 %
2,30	200 m.	79 %
3,07	250 m.	79 %
3,44	300 m.	79 %
4,19	350 m.	78 %
4,53	400 m.	79 %
5,26	450 m.	78 %
6,00	500 m.	78 %
	550 m.	
	600 m.	
	650 m.	
	700 m.	
	750 m.	
	800 m.	

Let tempo

Borgs Dyspnøskala (0-10)

FØR	0
EFTER	5

Gangdistance

500 m

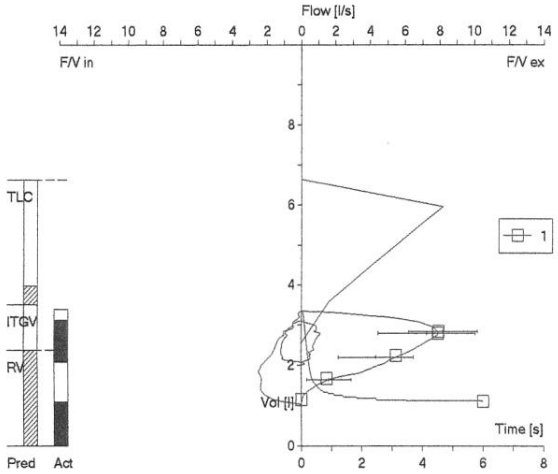
Mette Aukjær

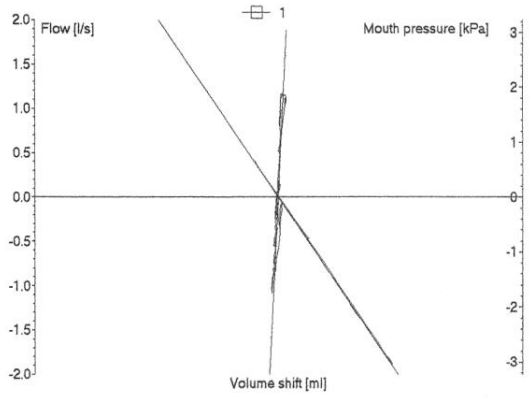
Odense Universitetshospital * Medicinsk afd. C * Lungemedicinsk/allergologisk sektion

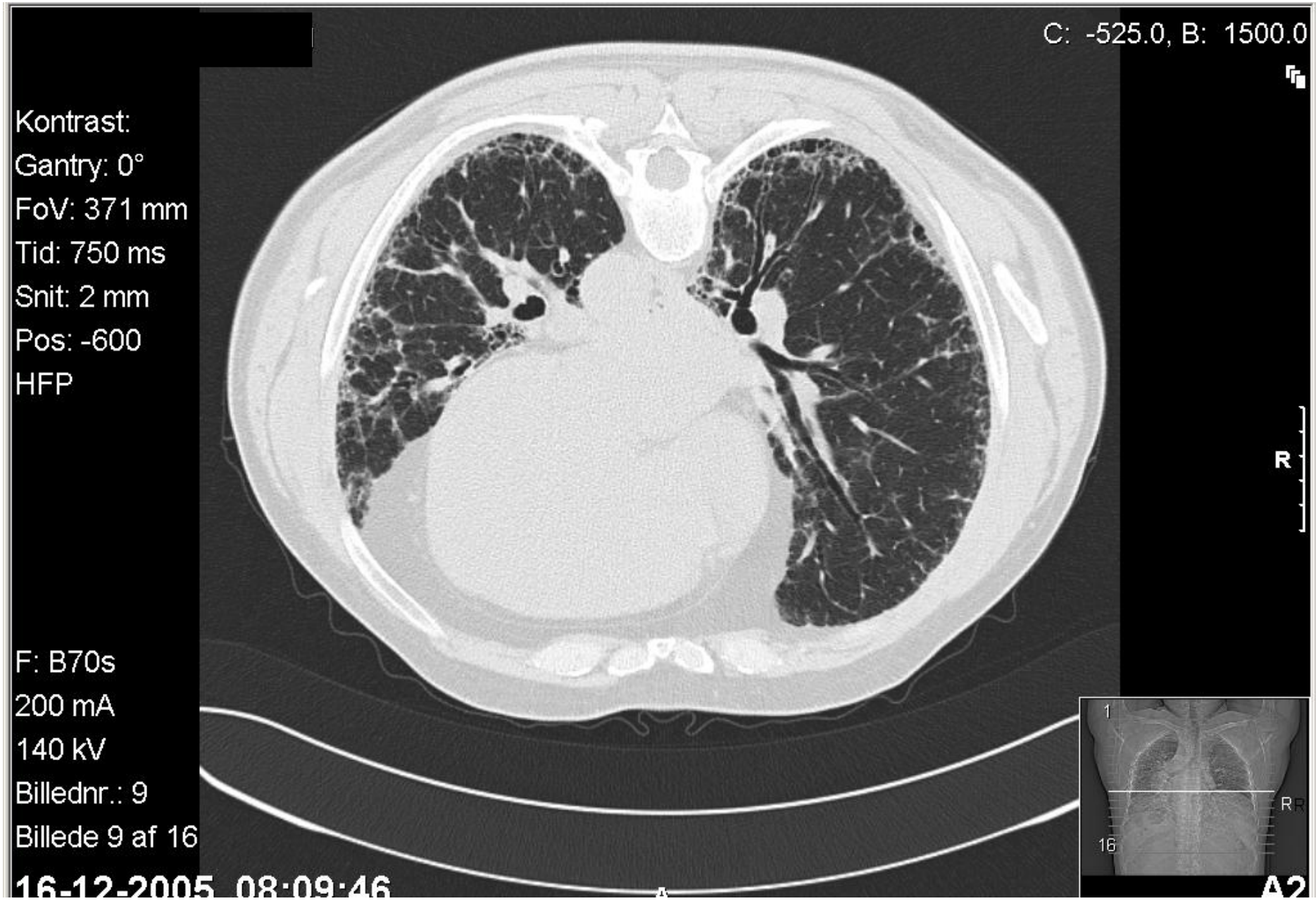
UDVIDET LUNGEFUNKTIONSUNDERSØGELSE

Identification:
First Name:
Last Name:
Height: 174,0 cm
Rel. Weight: 92 %
Operator:
Physician: --
Last Test: 1

	Pred	Act1	Act2	Act3	(A1/P) %	(Best/Pred)
Date		07-07-2006				
ATS-ok		0.00				
FEV 1	3.22	2.08			64.6	
FVC	4.10	2.24			54.8	
FEV1%F		92.71				
FIV1		1.72				
MIF		0.68				
MEF		0.44				
PIF		2.47				
AIN						
TLC	6.82	3.41			50.0	
RV	2.39	1.09			45.6	
RV%TLC	37.75	31.97			84.7	
Hb		15.40				
TLCOc	9.28	2.97			32.0	
KCOc	1.36	0.98			72.4	



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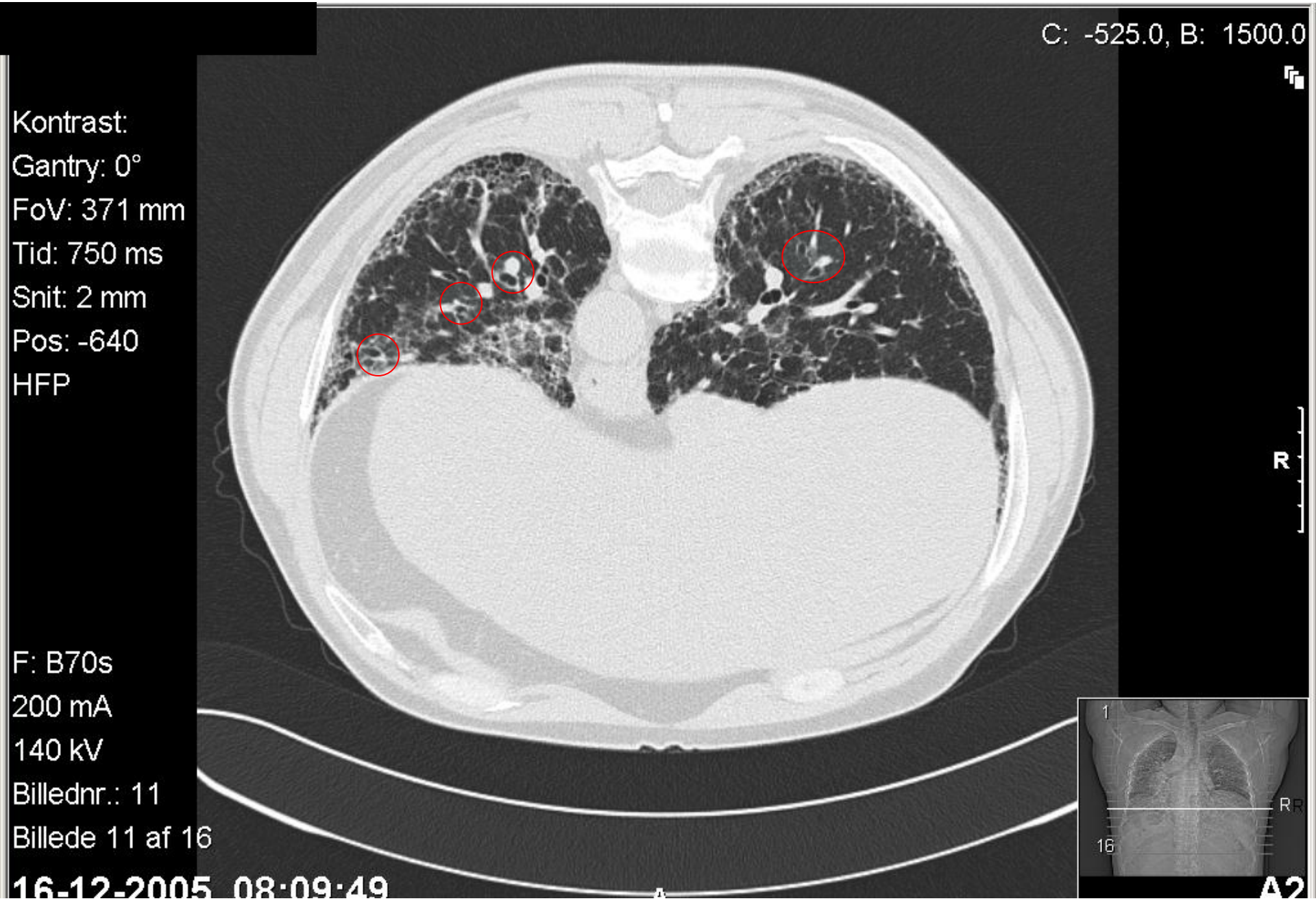
HRCT skan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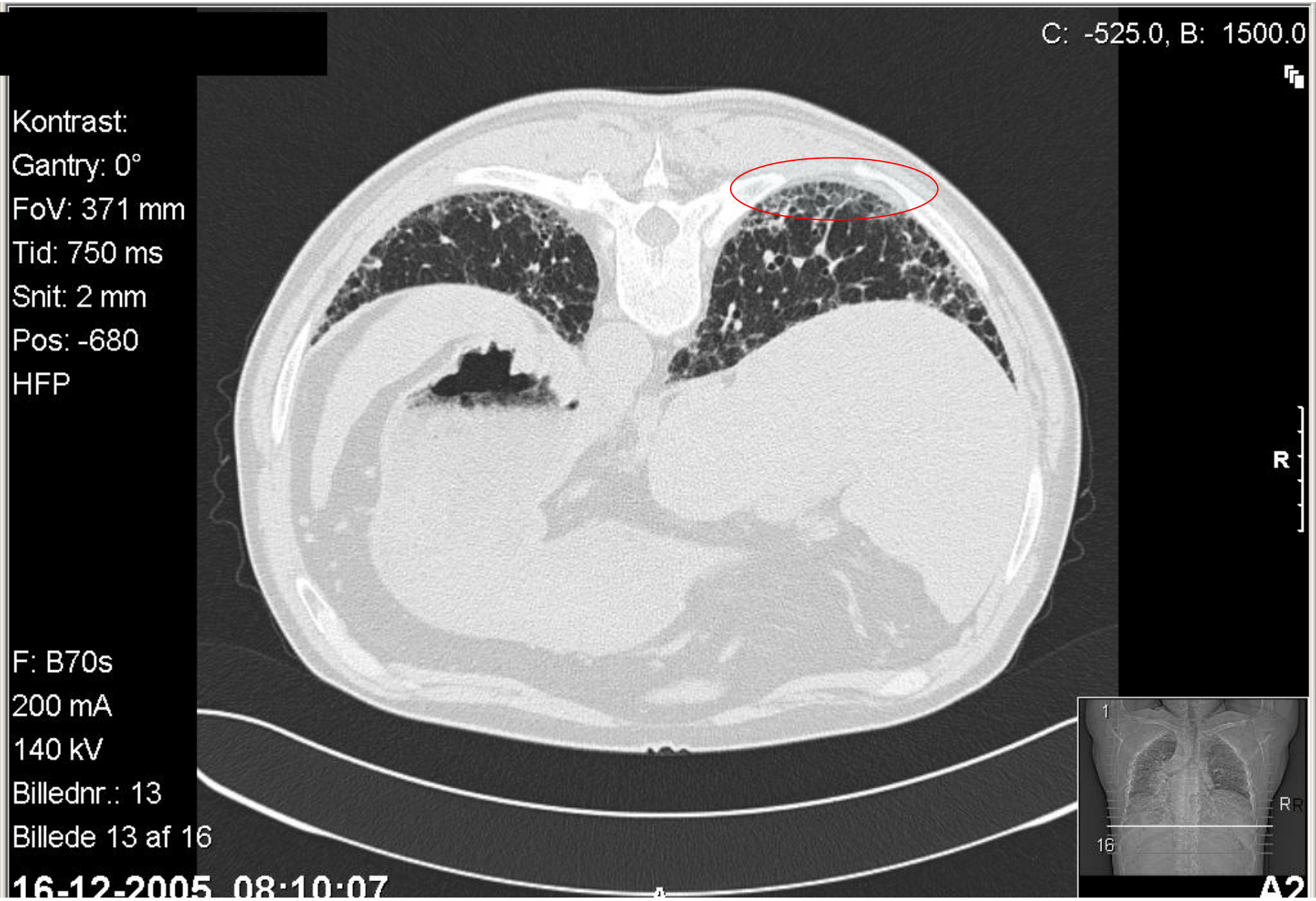
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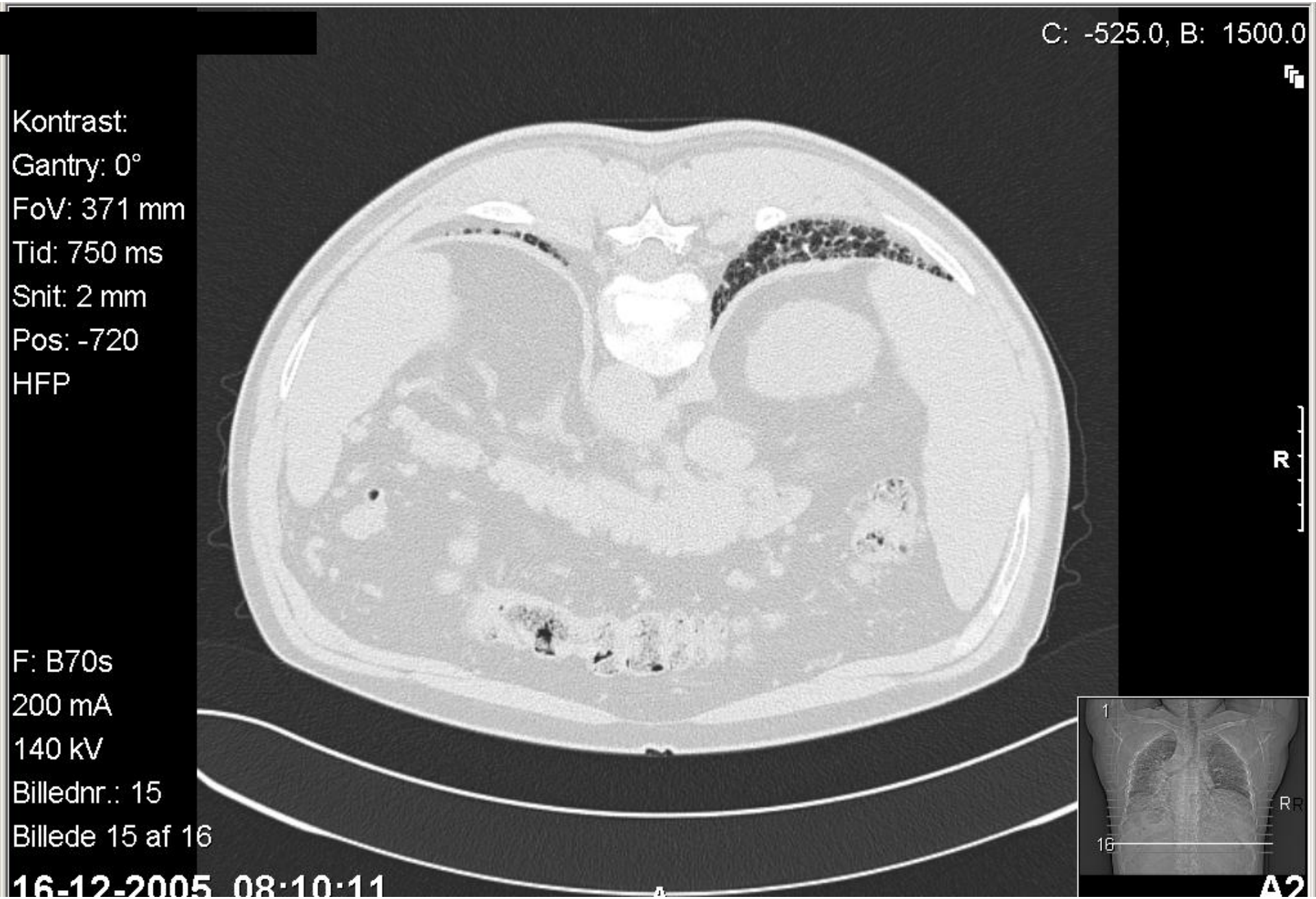
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Bronchial lavage for flow cytometri is done and shows total cellenumber of 26 mio., distribution is 66% makrofags, 2% lymfocyttes, 32% granulocyttes and of those 28% is neutrofile og 4% er eosinofile. .

Biospsy is suboptimal material very few parts of interstitielt lung tissue, nor enough to make a diagnosis . But signs of few active cells and some fibrosis. .

Resume:61 årig man, subjektive progression over 2 years, clubbing and vencrosound on stetoscopy; ANA positiv, 6 min walk sat. fra 91-78%, TLC 50%, diffusion: 32%; bronko. Shows inflammation with fibrosis og HRCT honny combing , tractions bronkieectasiers primary in the lowest lung part

Diagnosis ?

1. Patient has restriktive lung disease
2. Patient has DPLD but the excact diagnose is uncertain
3. Patient has allergic alveolis
4. Patient has sarcoidosis
5. Patienten has asthma
6. Patienten has COPD

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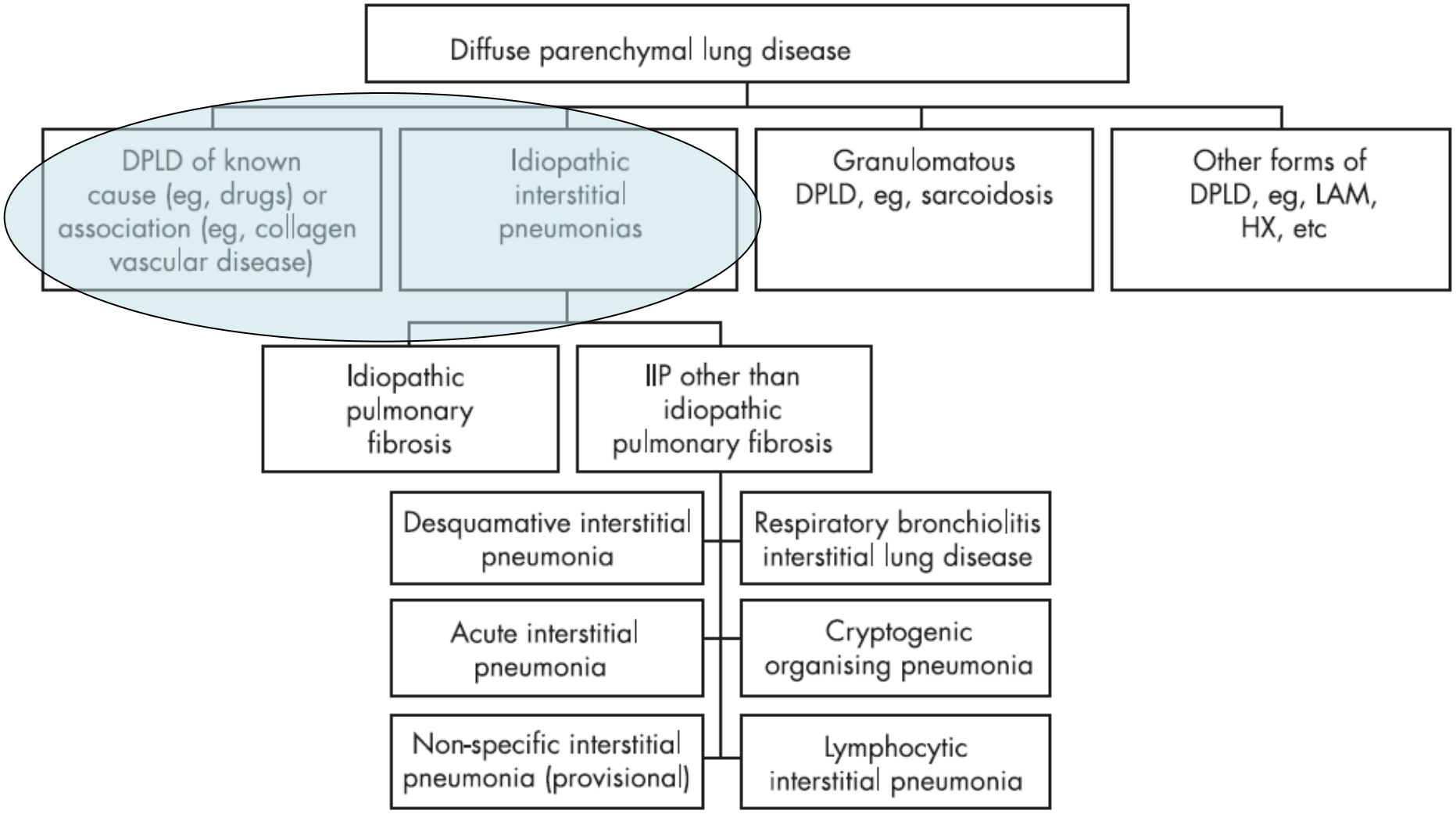
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Non-specific interstitial pneumonia (provisional)
Lymphocytic interstitial pneumonia

- Patienten is offered VATS mhp lung biospy to gain a more specific diagnosis

He refuses initially !!!!

Diffuse Parenchymal Lung Disease (DPLD)

Incidence: 1 in ~ 3500

– pulmonary practice ~ 15% of patients

Usually a subacute or chronic clinical presentation with a slowly progressive course
- – Exertional dyspnea
- – Nonproductive cough
- – Hypoxemia
- – Restrictive pulmonary function

Many heterogeneous clinicopathologic entities

– Etiology may be known (1/3) or unknown (2/3)

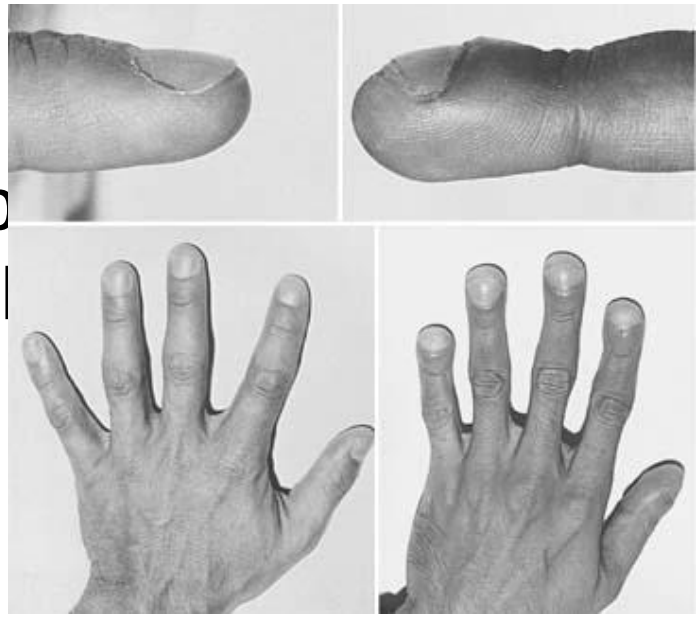
What do we see ?
A-gas

- On exercise PaO_2 decreases dramatically.
- Arterial PaO_2 are reduced, pH normal.
- Physiologic dead space and physiologic shunt and VQ mismatch are increased.
- Diffuse impairment contributes to hypoxemia on exercise.
- There is marked reduction in diffusing capacity due to thickening of blood gas barrier and VQ mismatch.

Clinical presentation

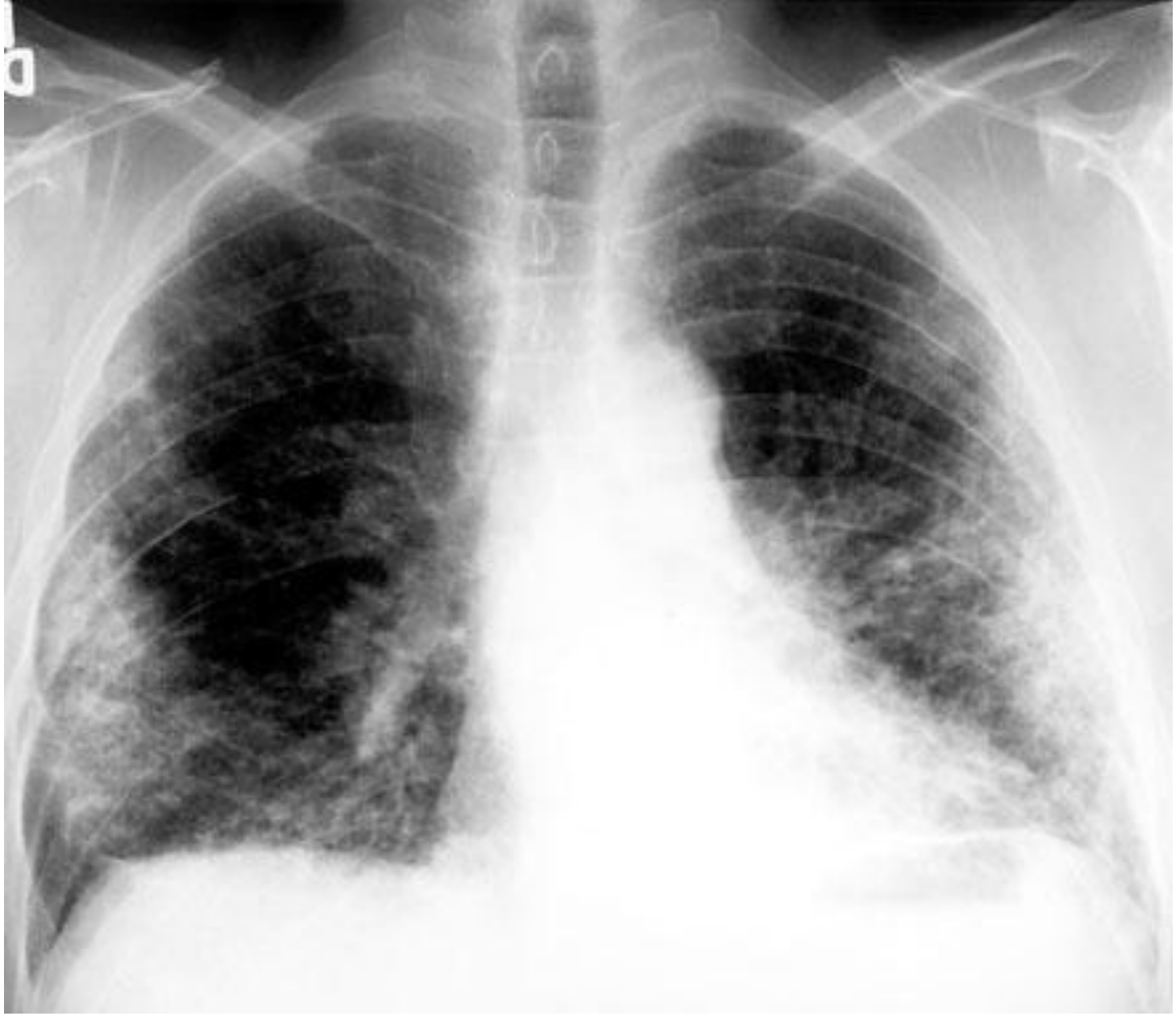
Subacute or chronic onset, Exertional dyspnea, nonproductive cough,
Constitutional Symptoms

- Tachypnea, digital clubb
- Inspiratory “velcro” crac
- Hypoxemia,
- cor pulmonale



Abnormal chest x-ray
– Reticular, reticulo-nodular patterns
– Distribution (bases, periphery)
– Honeycombing
– Ground-glass pattern (HRCT criteria, not on CXR)
Pulmonary symptoms associated with another disease, such as a connective tissue disease

Lung function abnormalities



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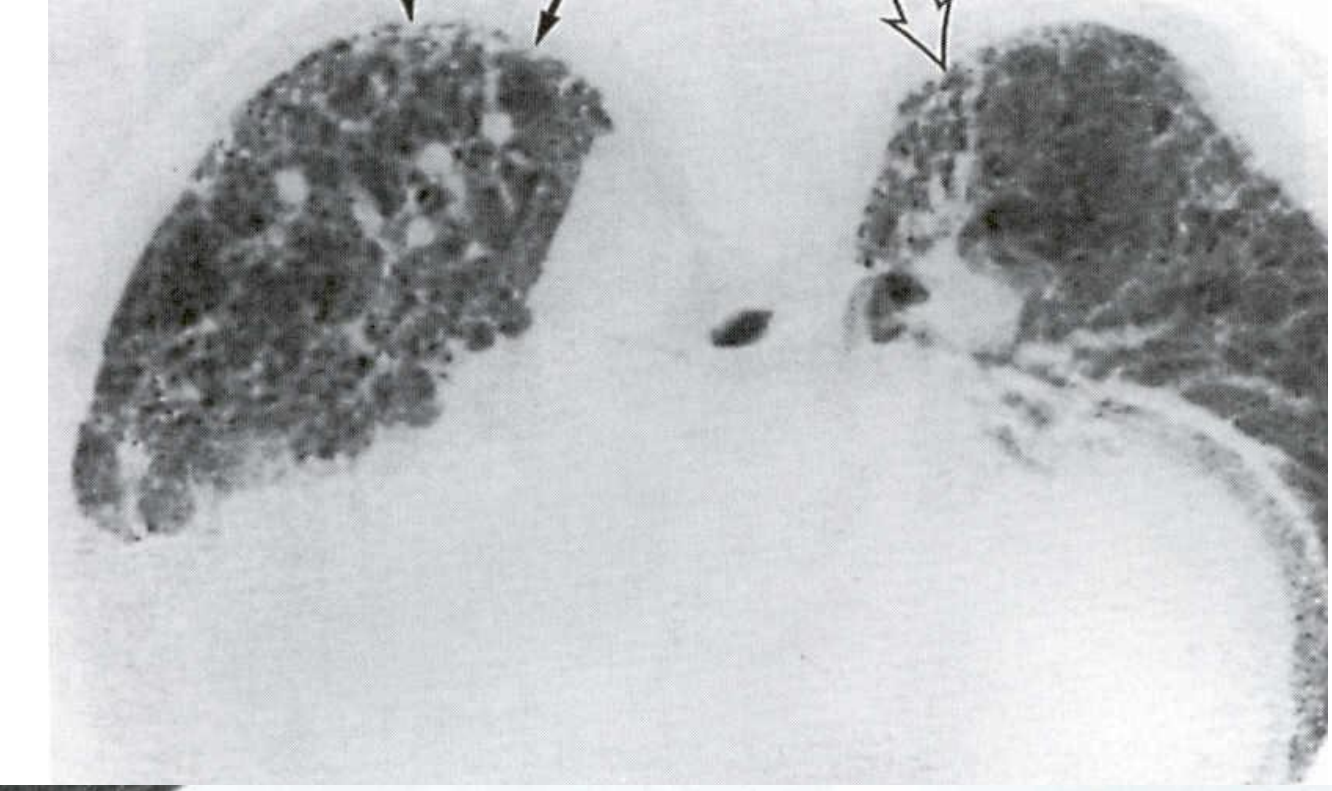
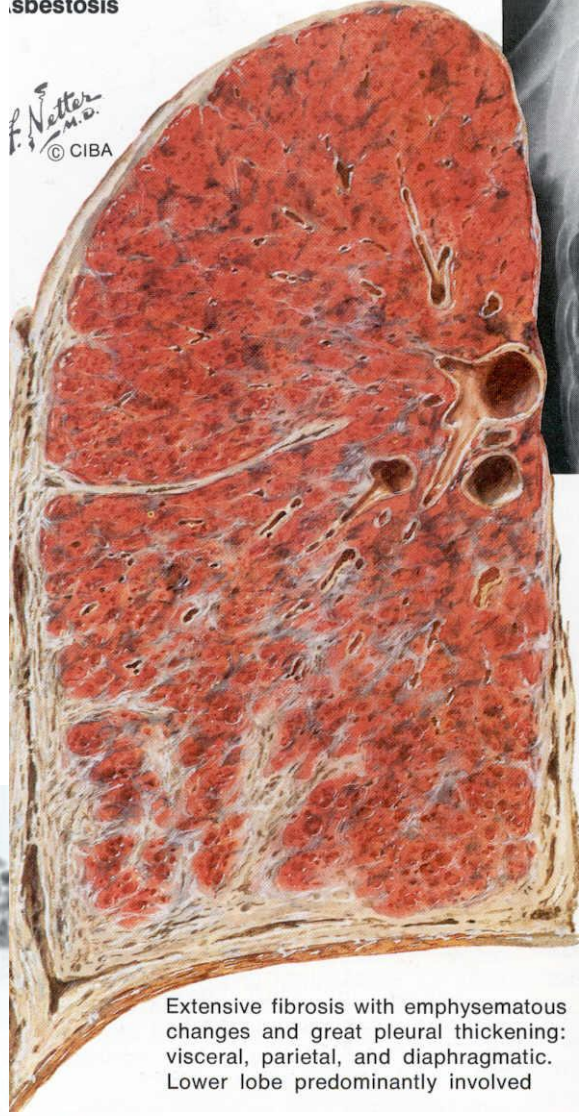
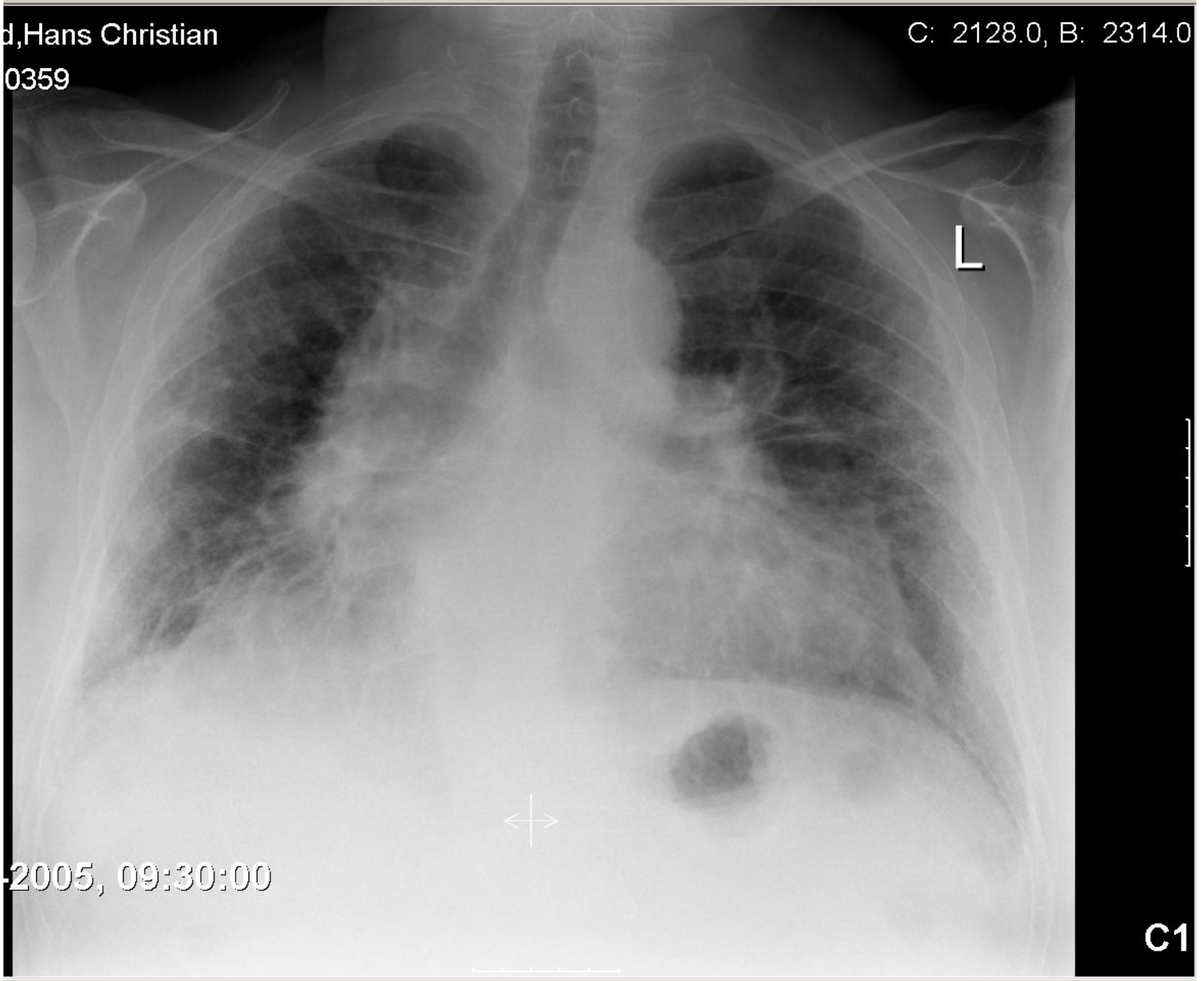




Extensive fibrosis with emphysematous changes and great pleural thickening: visceral, parietal, and diaphragmatic. Lower lobe predominantly involved

- One to see !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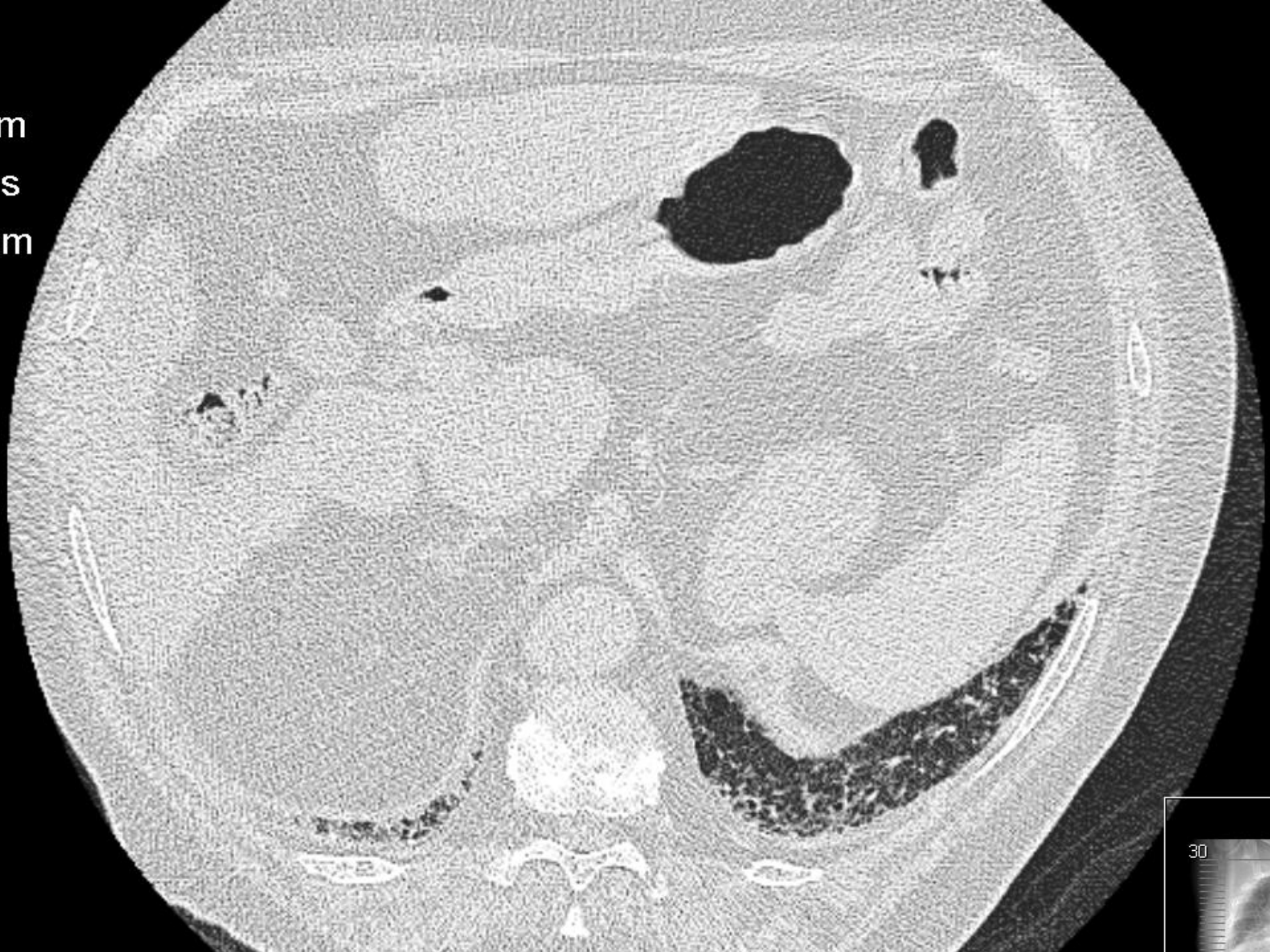
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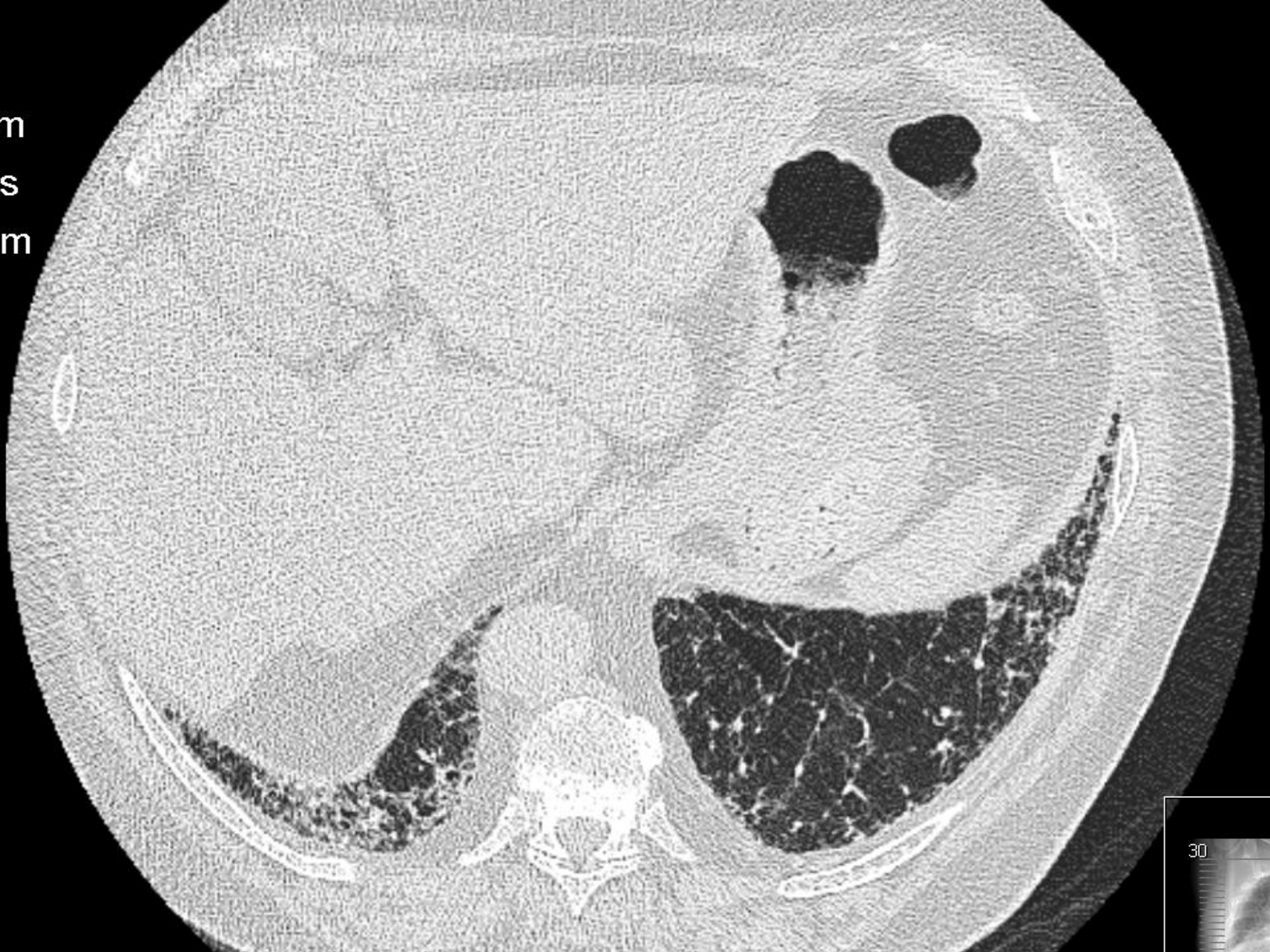
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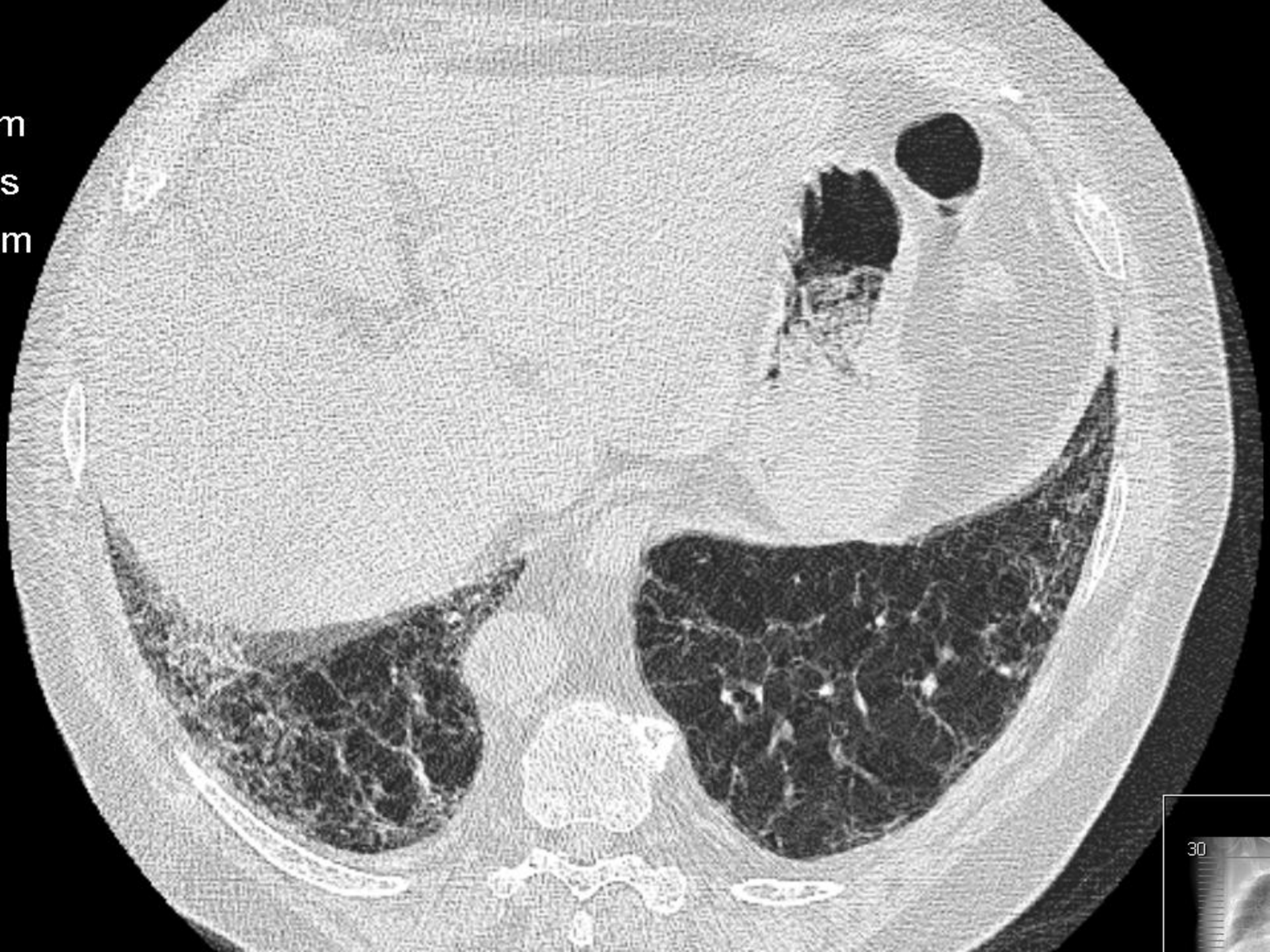
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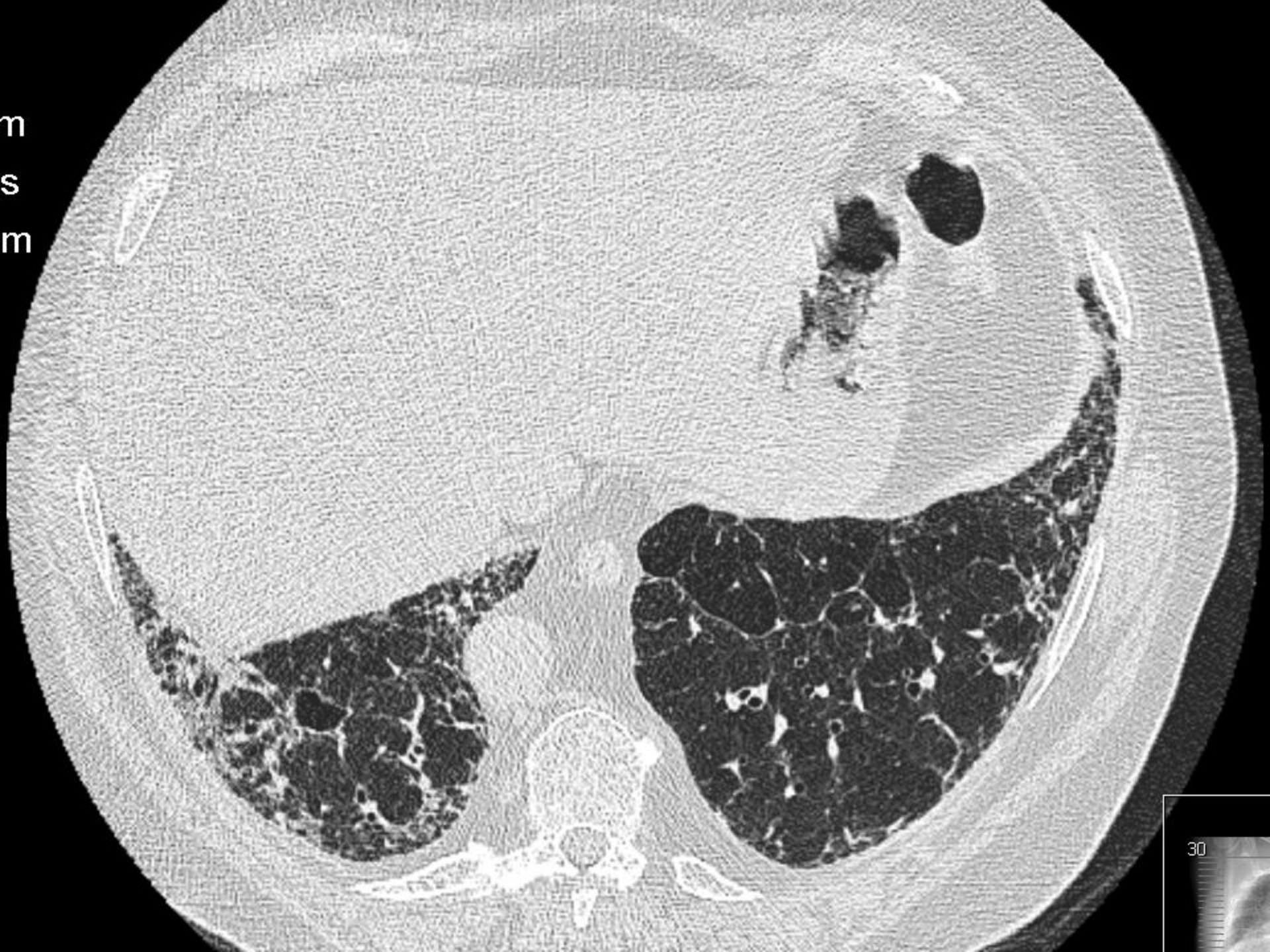
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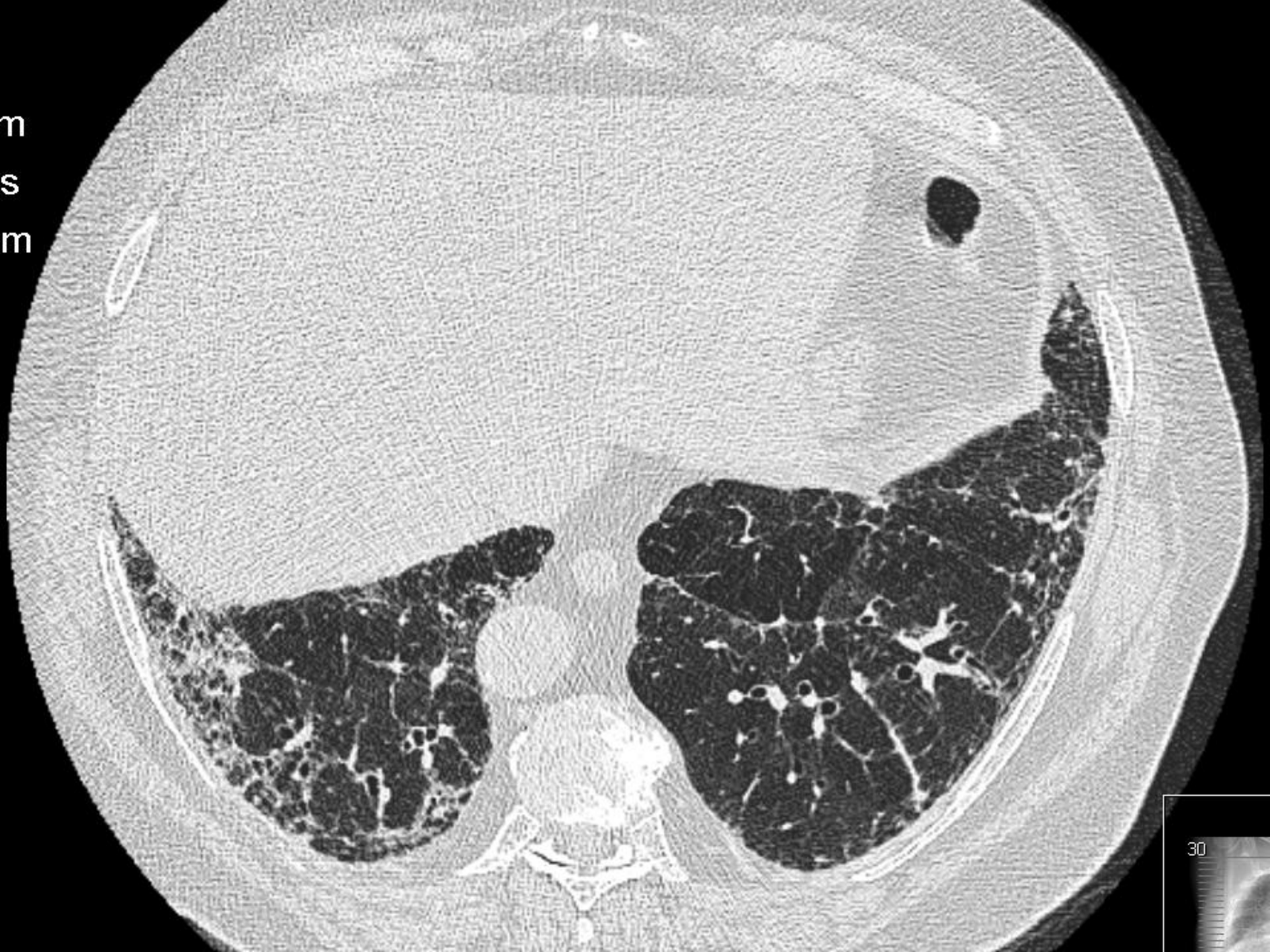
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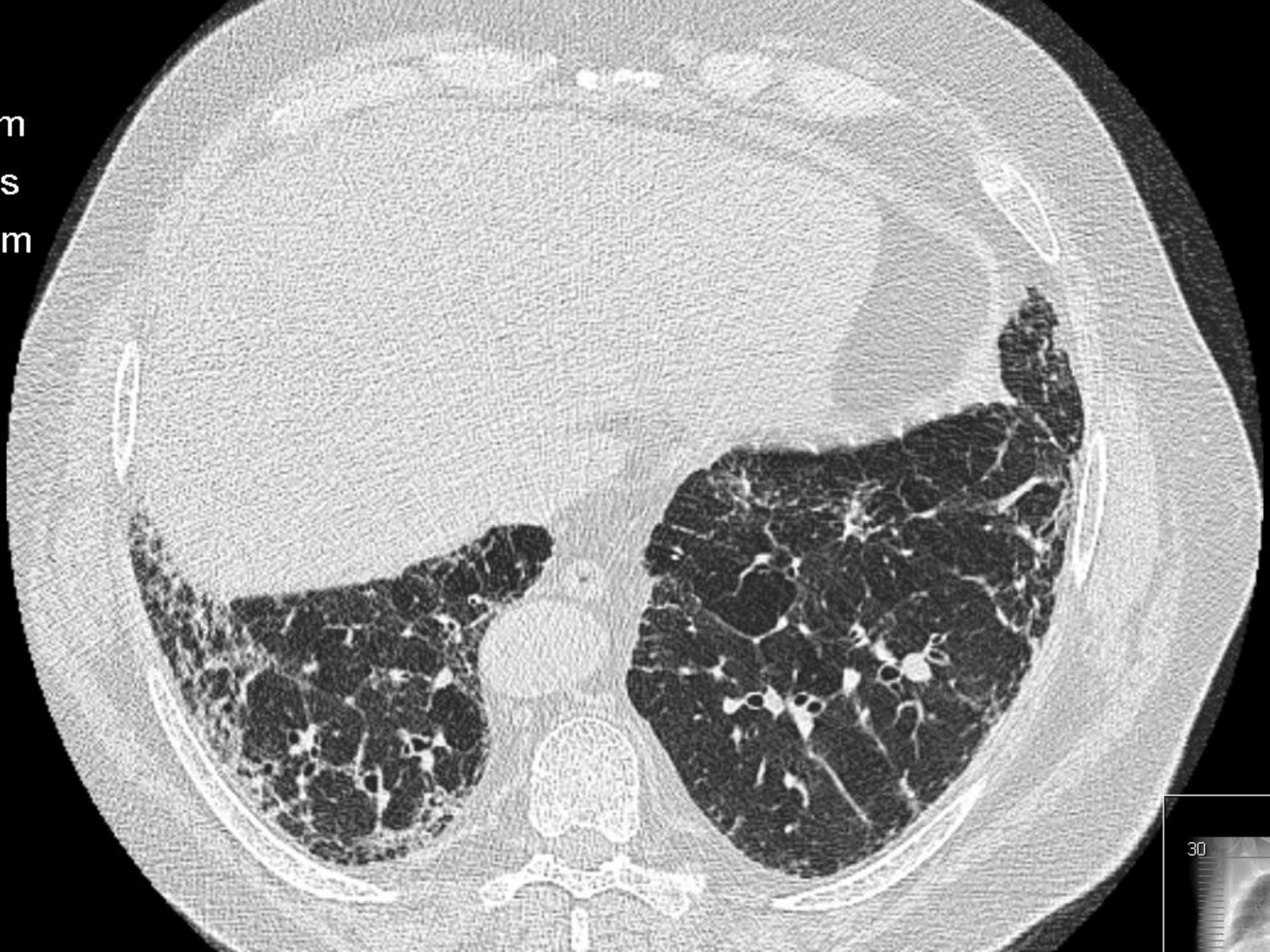
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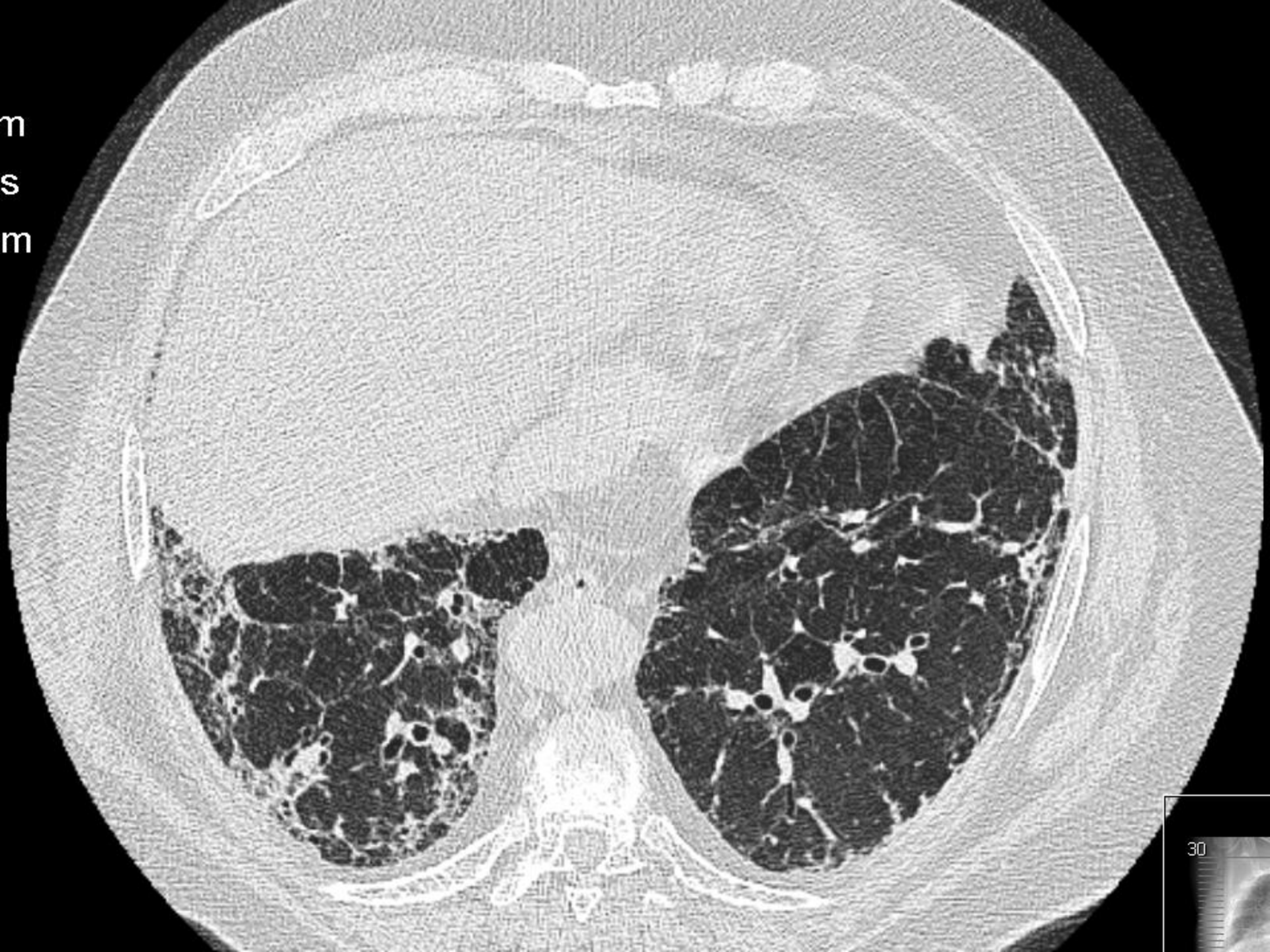
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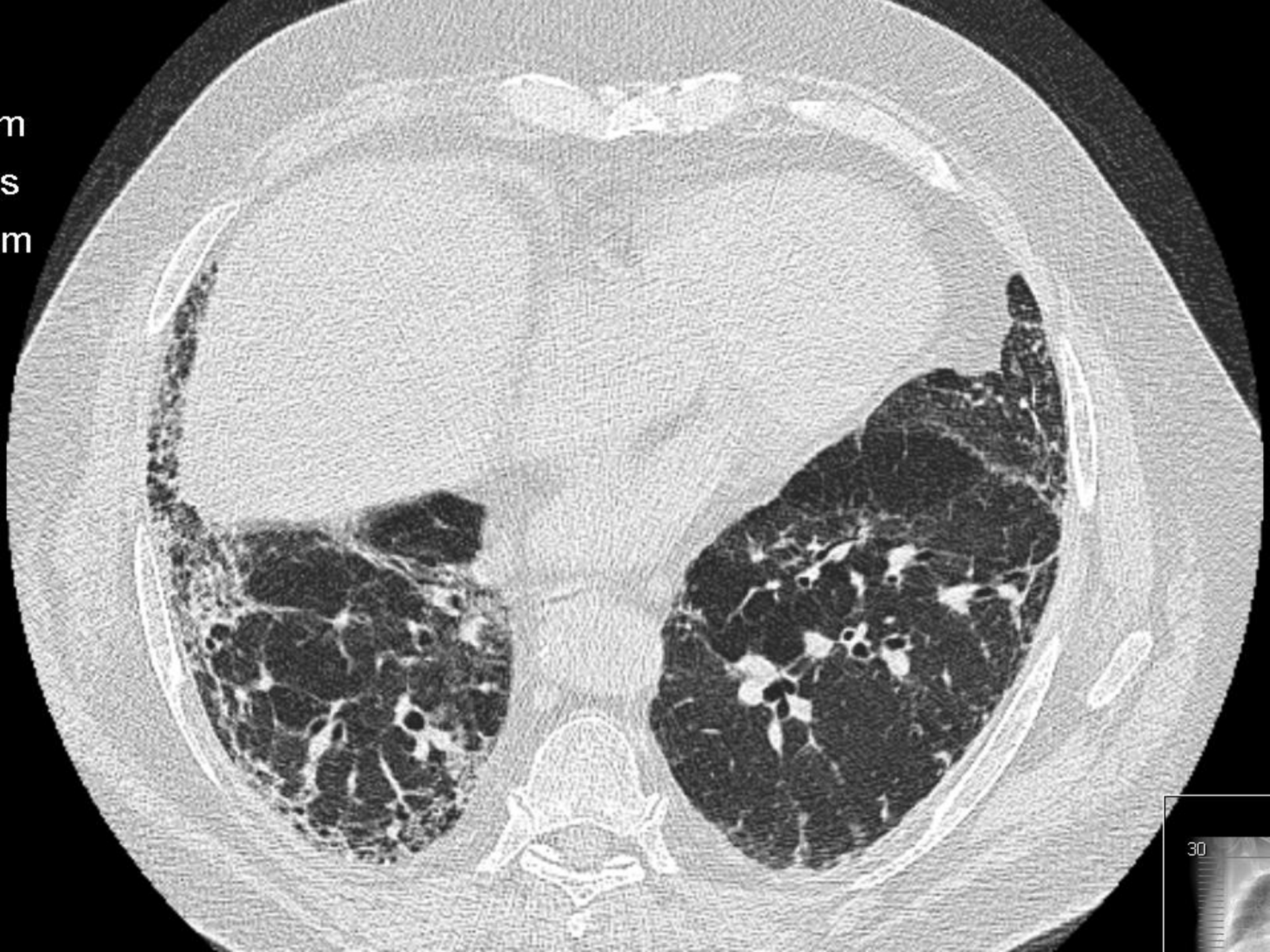
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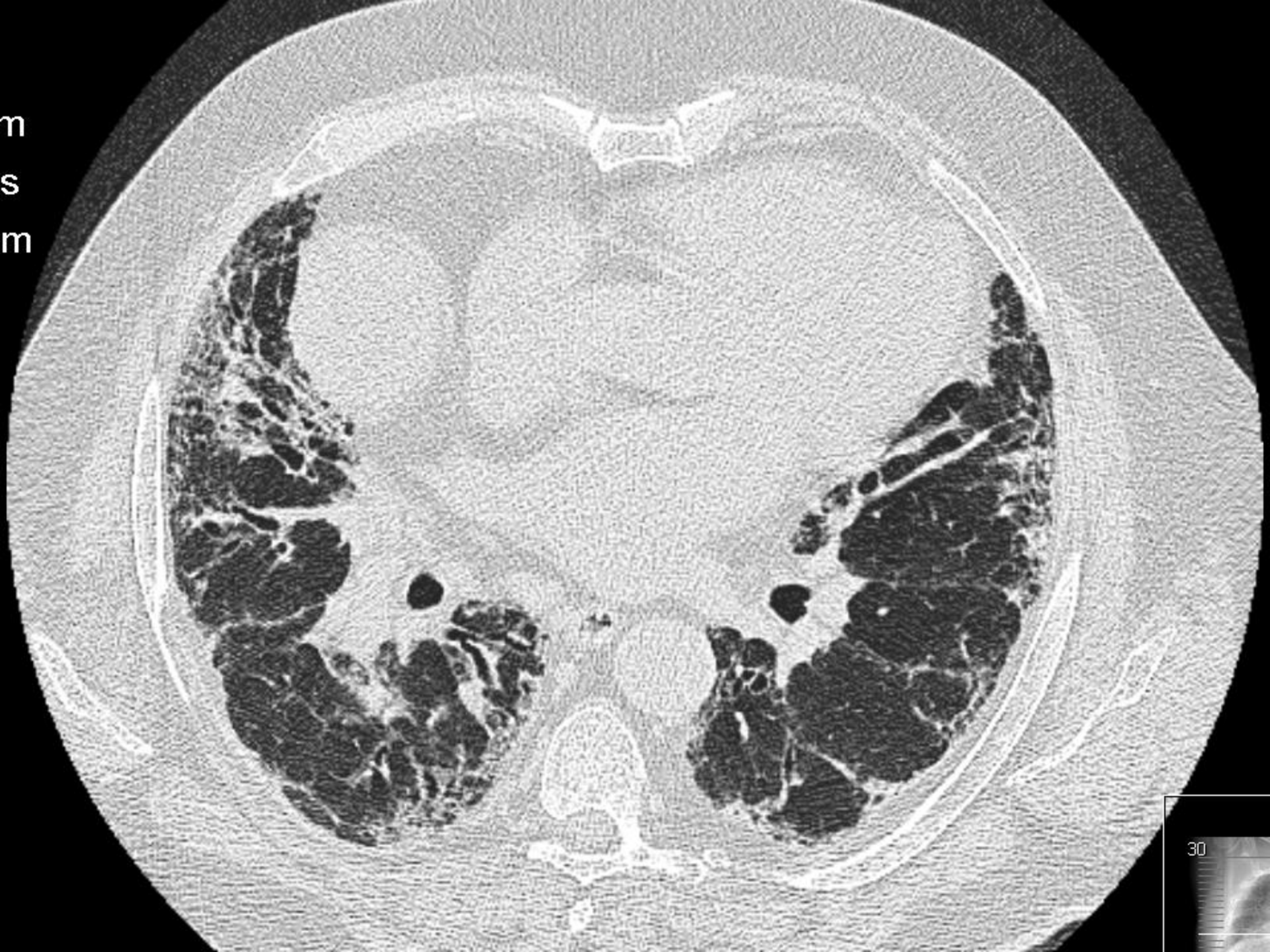
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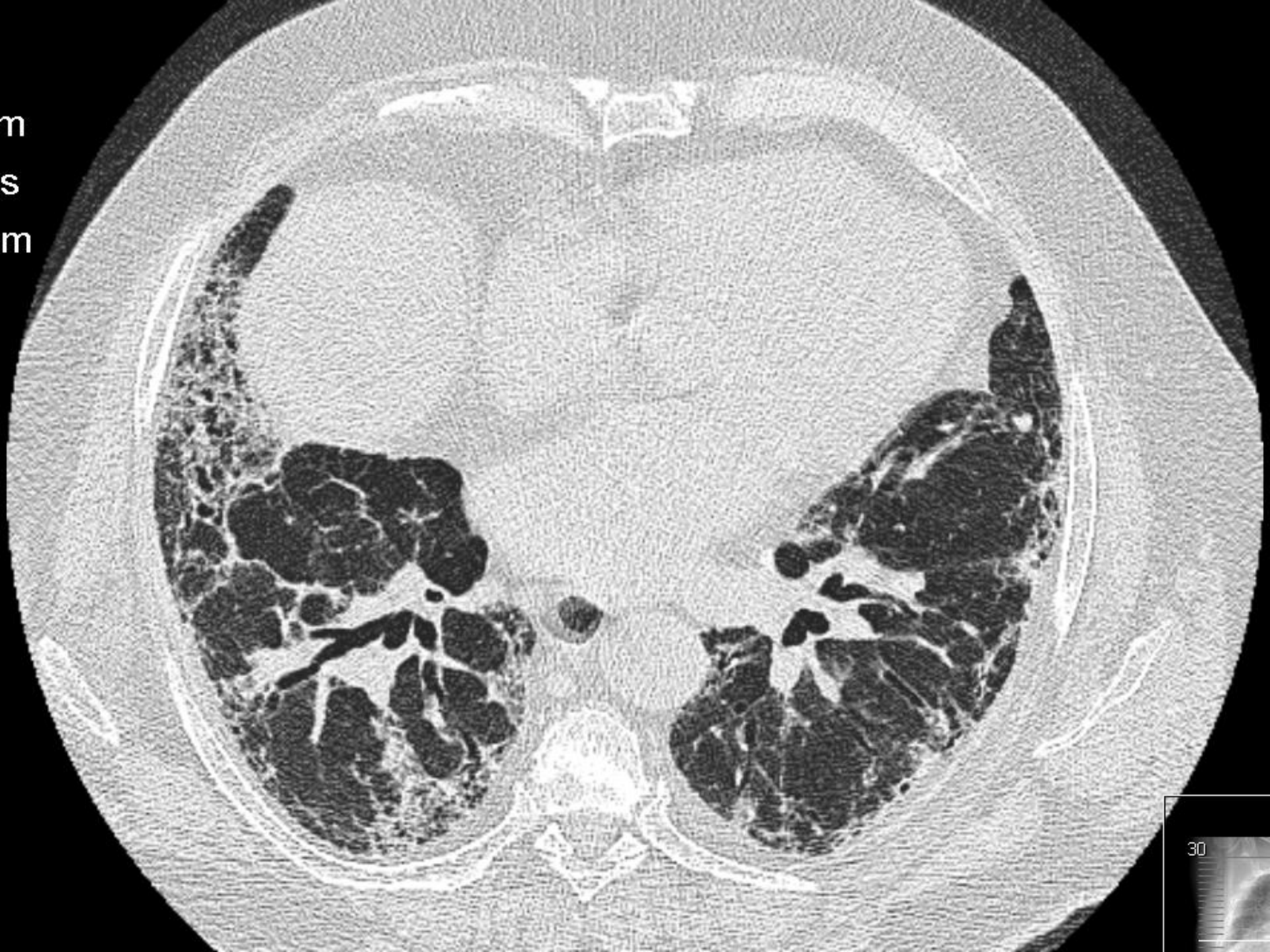
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- 03.04.08
- HRCT shows bronkieektasies in both sides from apex to basis. Lower subpleurale cysts are seen and clasic honney combing seen in idiopatisk lung fibrosis. Apikalt findes øget interstitiel tegning.
- RD: Lung fibrosis with bronkieektasies

biopsi was done : B: firekantet perfusionsfikseret lungeresektat med stapler på to kanten. Resektatet måler 48 x 33 x 8 mm. Vævet fremtræder lidt uregelmæssigt med diskrete fibrotiske områder. Alt med i 6 kaps. undtagen den staplede kant./jyc

Undersøger: KEO/pke

MIKROSKOPI BESKRIVELSE:

A-B: Alt materialet er indstøbt og undersøgt i histologiske snit, hvor der ses heterogent lungevæv. I samme synsfelt forekommer normalt lungevæv, fibrose med cystedannelser samt område med kronisk inflammation. Der er desuden en del sekretstagnation til dilaterede luftveje. De inflammatoriske områder viser beskeden inflammatorisk aktivitet, overvejende med lymfocytter, men også spredt forekomst af eosinofile granulocytter. Der er regeneration

PATIENT: 280639-0359

med fibroblastiske foci. I perifere luftveje ses bronkial metaplasi. I områder med forandringer af mere kronisk karakter ses bindevæv samt glat muskelcellehyperplasi. Der er ikke granulomer, amyloid eller malignitet. Der er en del tykvæggede kar, hvilket opfattes som sekundært til lungeforandringerne.

conclusion :

good material shows fibroserende alveolitis (UIP). /dpa

Important

The process of achieving a diagnosis in a patient with interstitial lung diseases is dynamic, requiring close communication between clinician, radiologist, and pathologist.

2002 Jan 15;165(2):277-304

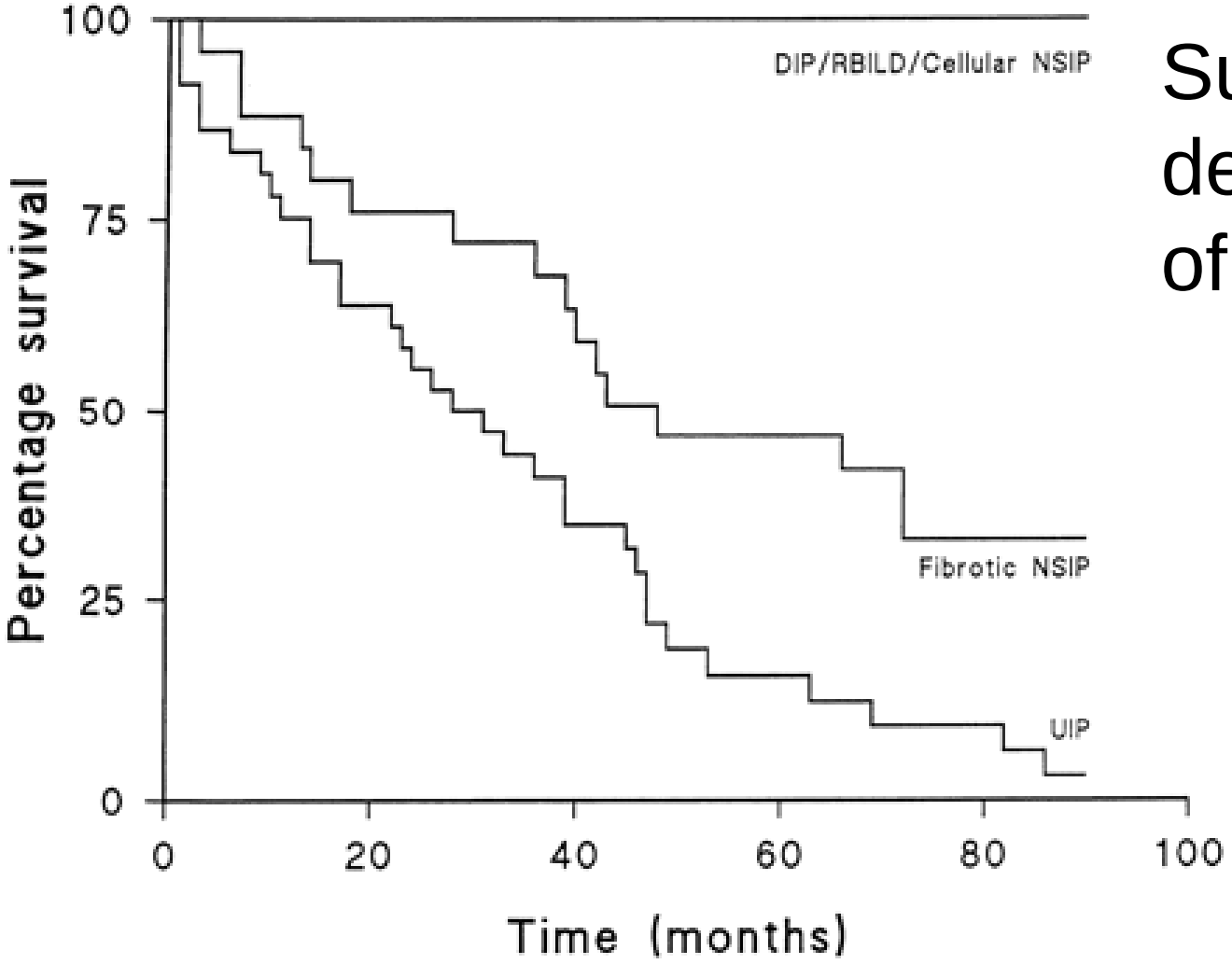
Diffuse Parenchymal Lung Disease

There is a single fundamental question which will determine management and prognosis:

Does this patient have idiopathic pulmonary fibrosis?

IPF is the clinical/radiologic equivalent of Usual Interstitial Pneumonia (UIP)

- Empiric treatment is discouraged until a firm diagnosis is established

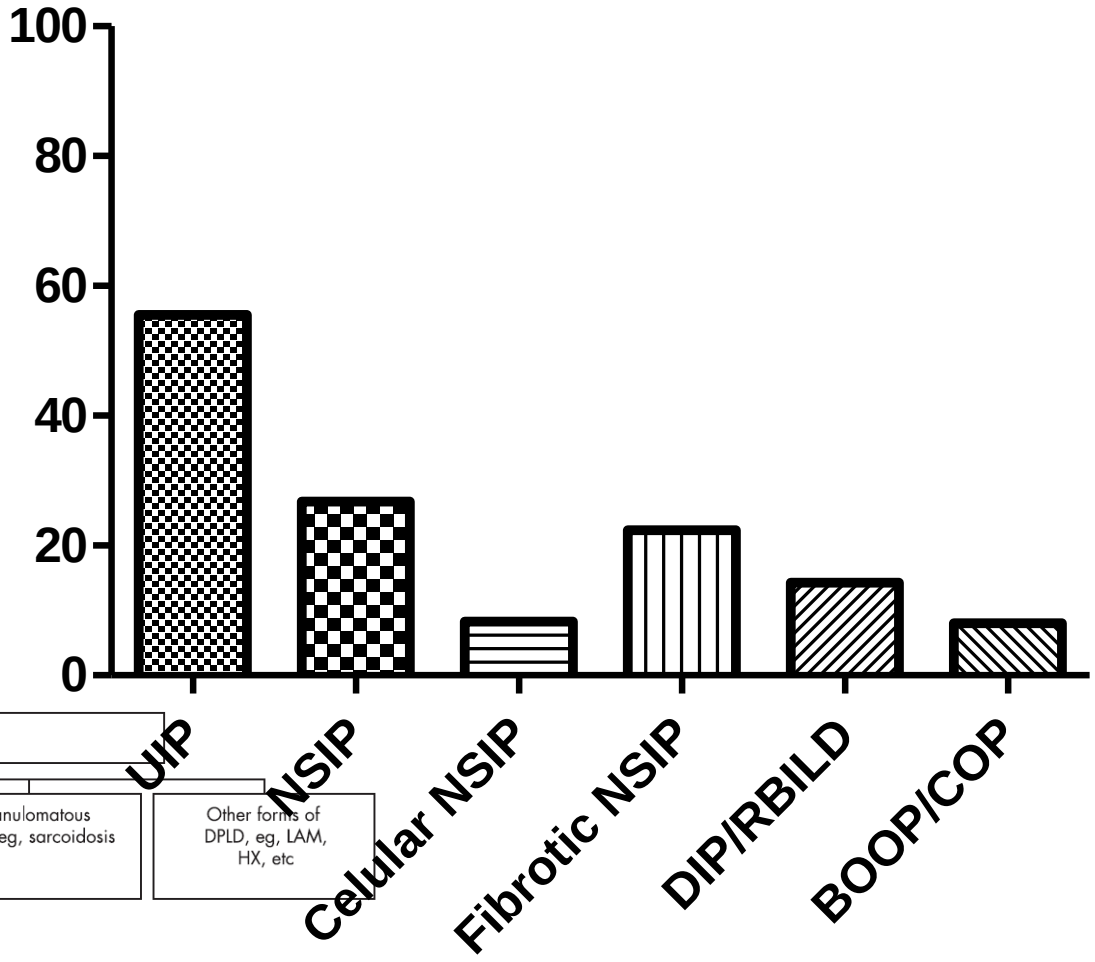


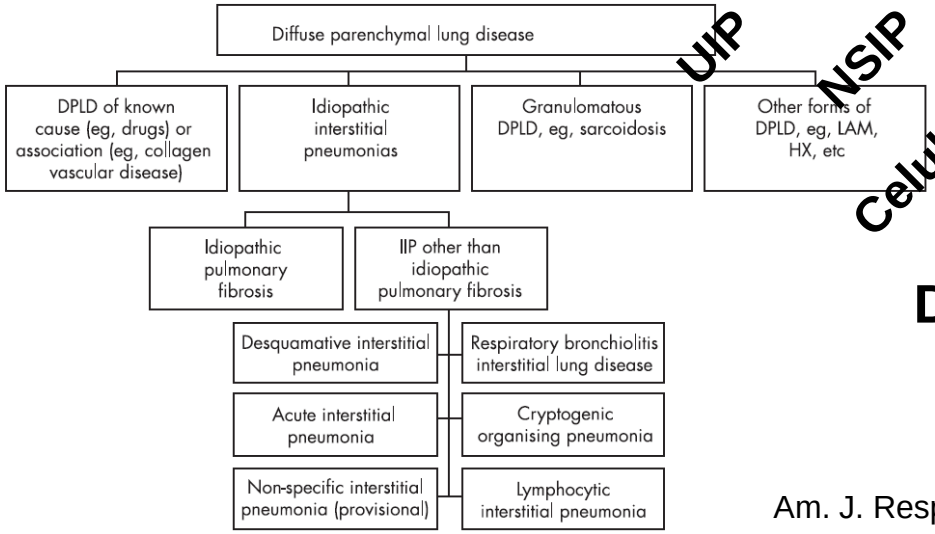
Survival of dependent of diagnosis !!

usual interstitial pneumonia (UIP), fibrotic nonspecific interstitial pneumonia (NSIP), and desquamative interstitial pneumonia (DIP)/respiratory bronchiolitis-associated interstitial lung disease (RBILD)/cellular NSIP

Am. J. Respir. Crit. Care Med., Volume 162, Number 6, Dec. 2000, 2213-2217

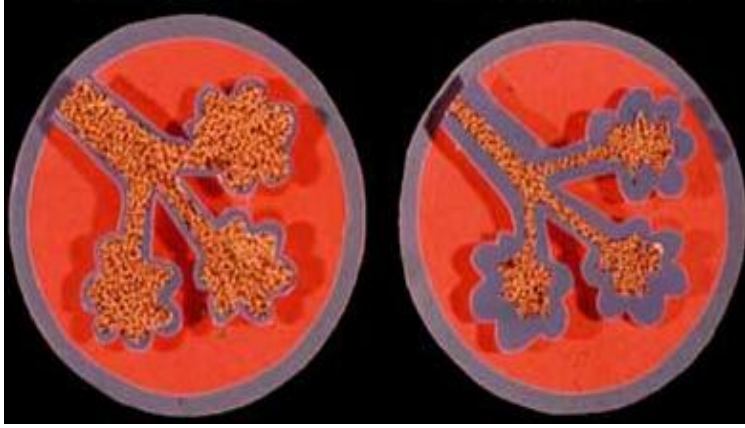
Prevalence of subgroup of interstitial pneumonia i 4 studies





Am. J. Respir. Crit. Care Med., Volume 162, Number 6, Dec. 2000, 2213-2217

Remember !



Restrictive lung diseases are defined by reduced total lung capacity, vital capacity and functional residual capacity, but with preserved air flow

Alteration in lung parenchyma, diseases of the pleura, chest wall or neuromuscular

Lung parenchyma:

Exertional dyspnea, nonproductive cough,Tachypnea, digital clubbing, Inspiratory “velcro” crackles, Hypoxemia, cor pulmonale

Does the patient have UIP ? – important due to treatment and prognosis.

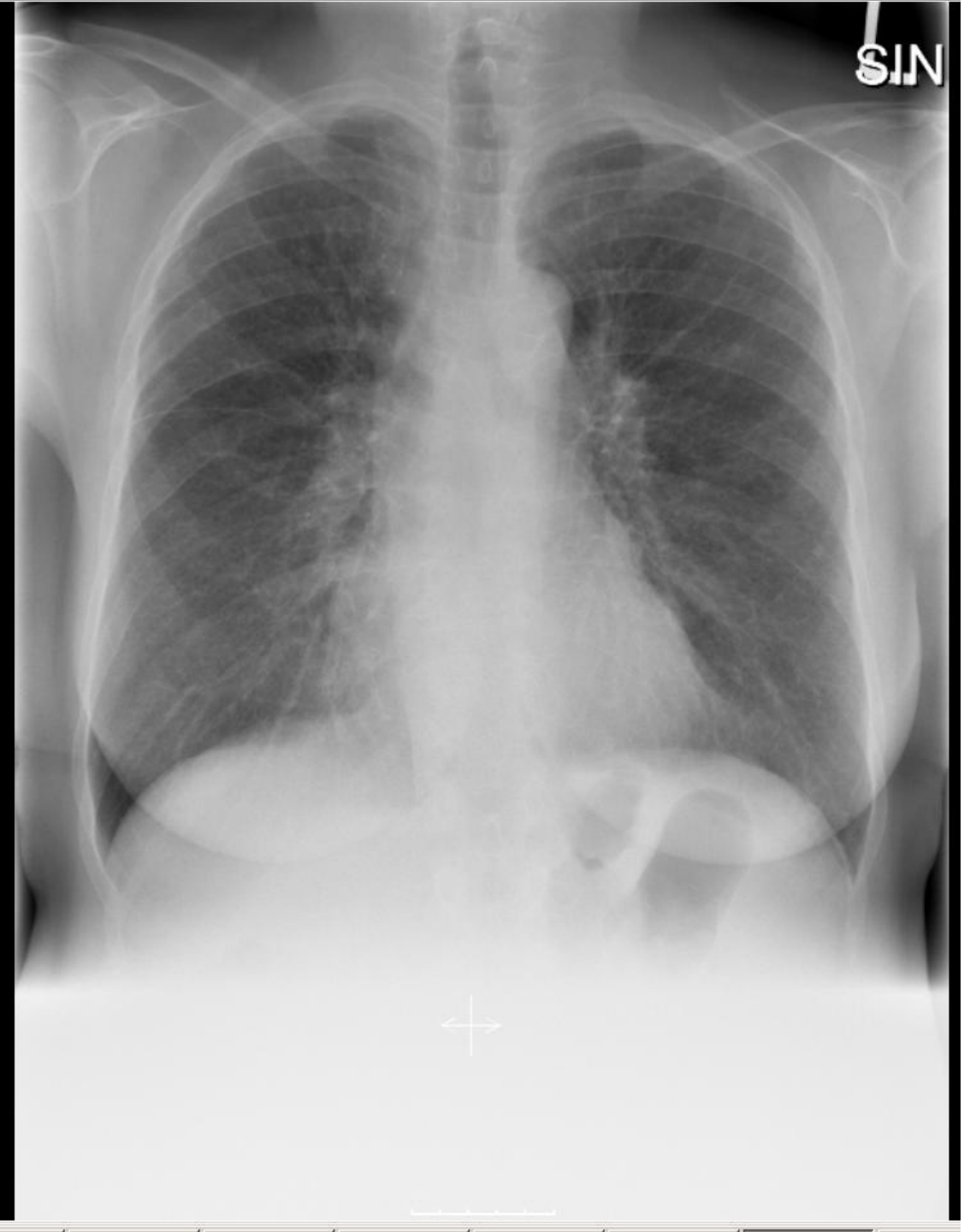
Myths !

- Smoking does NOT normally cause lung fibrosis
- However nothing without exceptions ..... Very rare lung diseases ……
- Smoking gives lungfibrosis…..???!!!

Smoking-Related Interstitial Lung Disease

- Respiratory bronchiolitis ILD (RBILD)
- Desquamative interstitial pneumonia (DIP)
- Pulmonary Langerhans' cell histiocytosis

Pigmented macrophages and inflammatory edema in the walls of the terminal and respiratory bronchioles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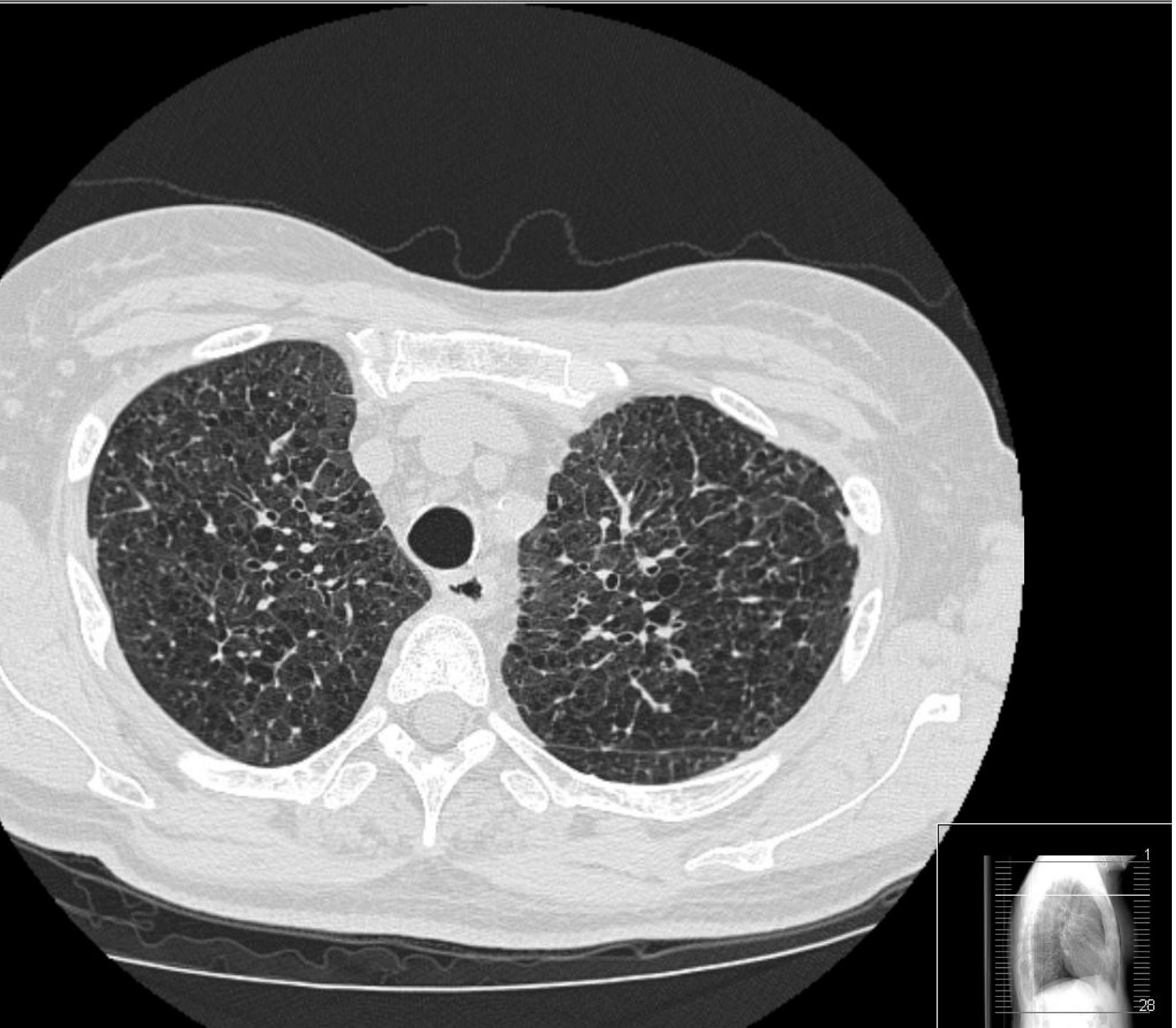
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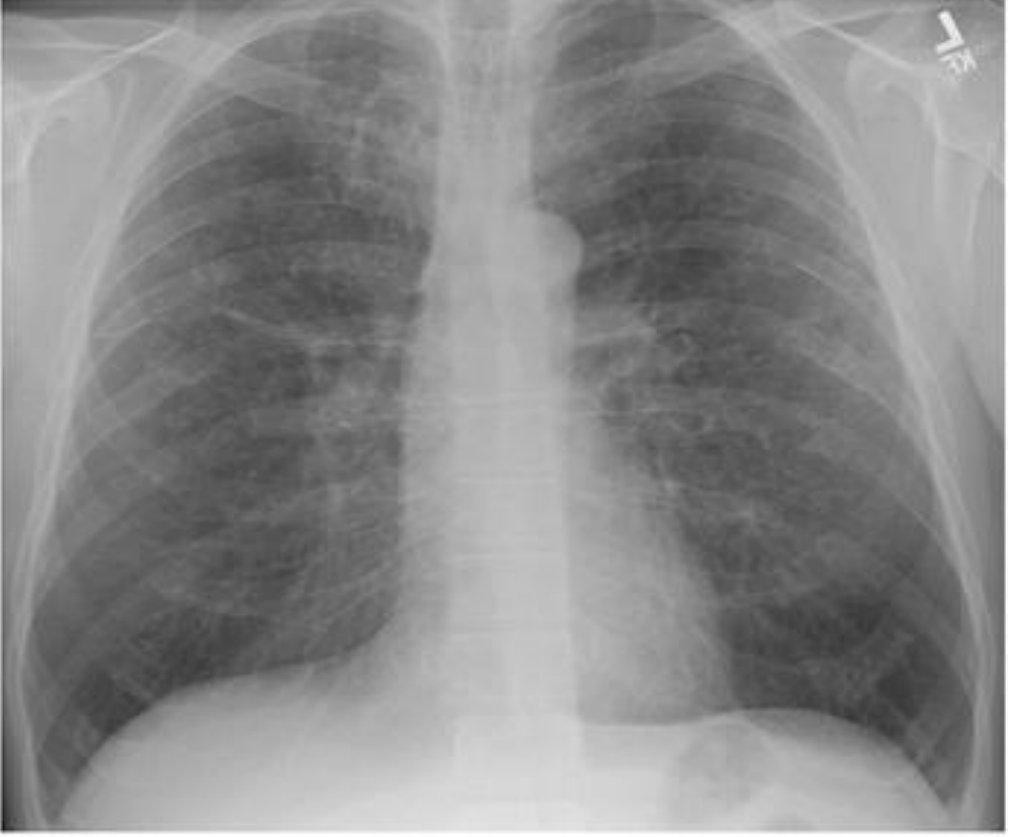
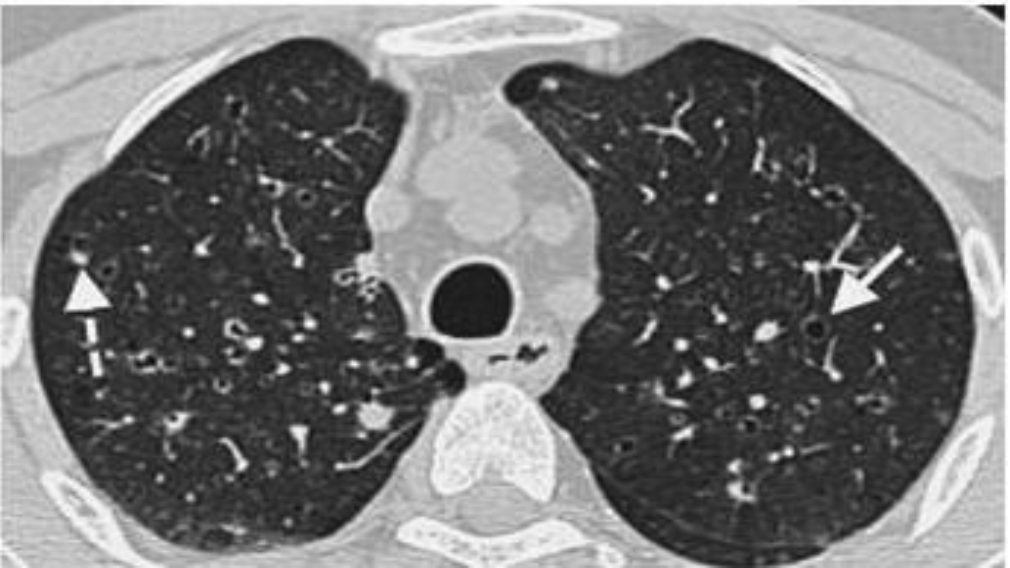




Langerhan cell histiocytosis. This 50-year-old man had a 30 pack-year history of cigarette smoking.

A: PA chest radiograph shows hyperinflation of the lungs and fine bilateral reticular ILD.

B: CT scan shows multiple cysts (solid arrow) and nodules (dashed arrow).

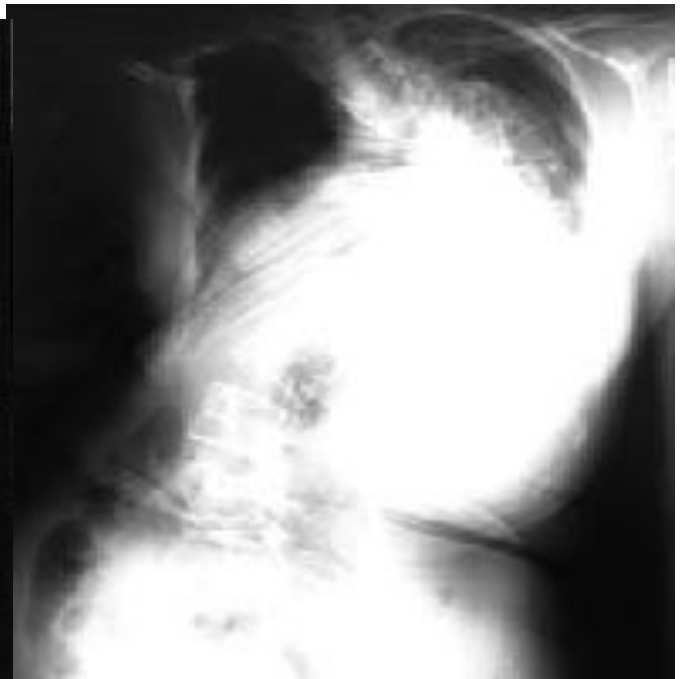

back | track
Back on Track
FØLG DIN ORDR
...interesting
TRACK & TRACE

Causes:
Restictive lung diseases

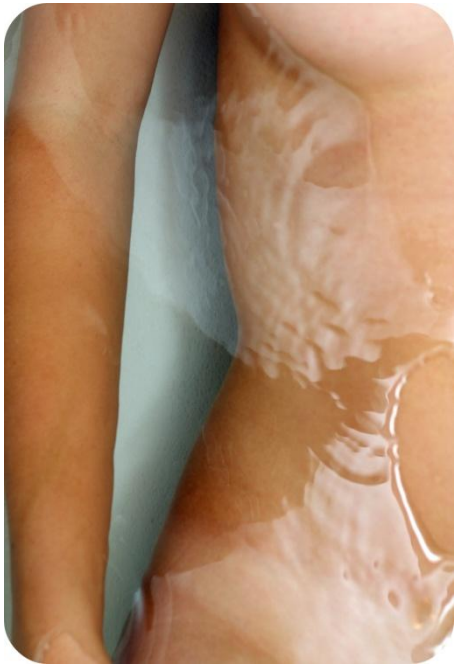
1. Changes in the lung parenkyma

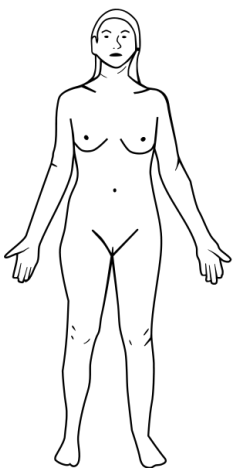
=Lung fibrosis

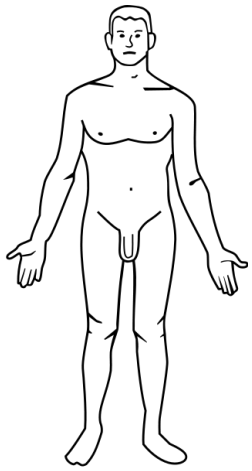
2. Diseases in the pleura, chest wall, muscles and nerves

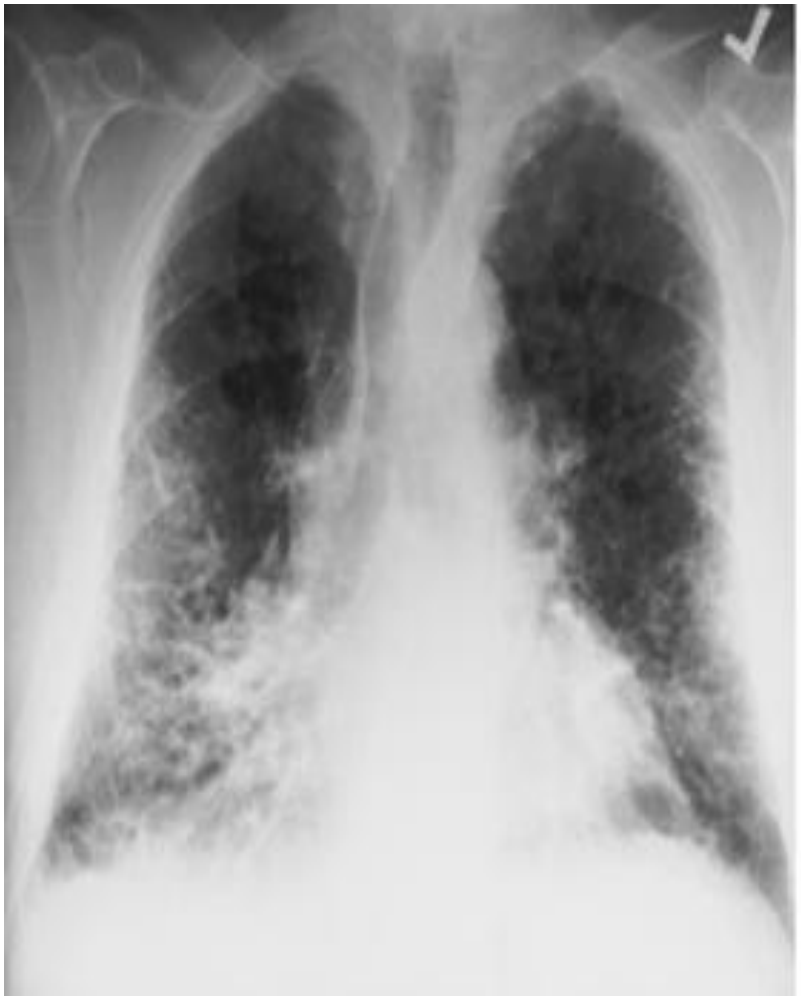


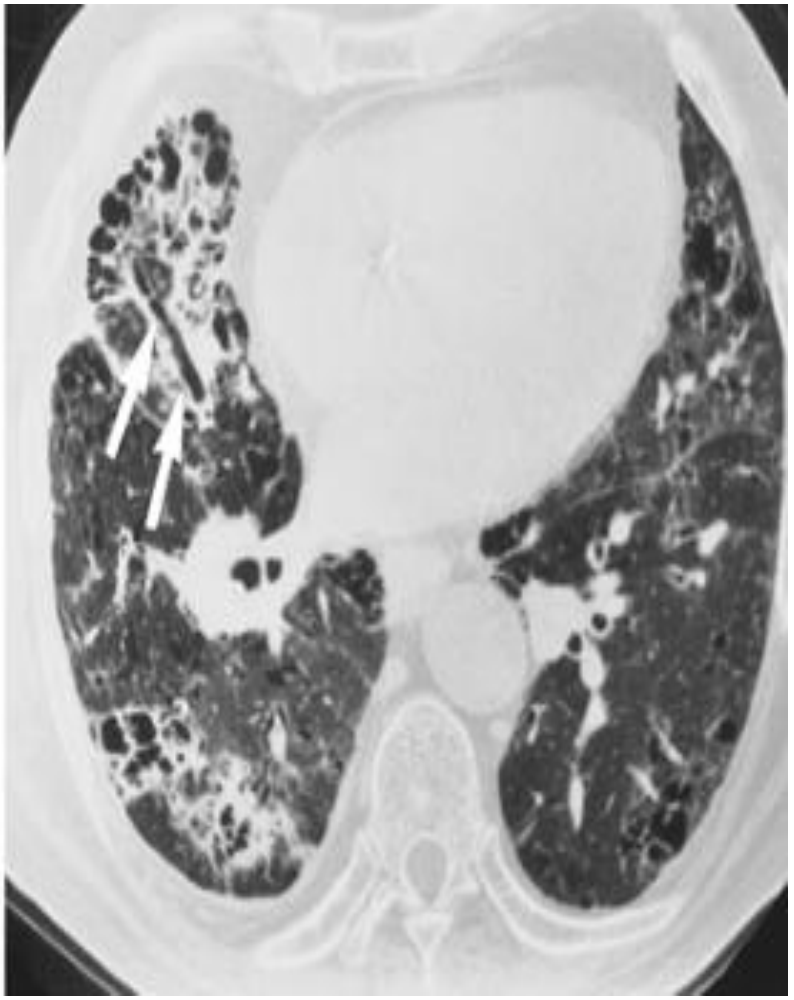
What happens in lung fibrosis ??





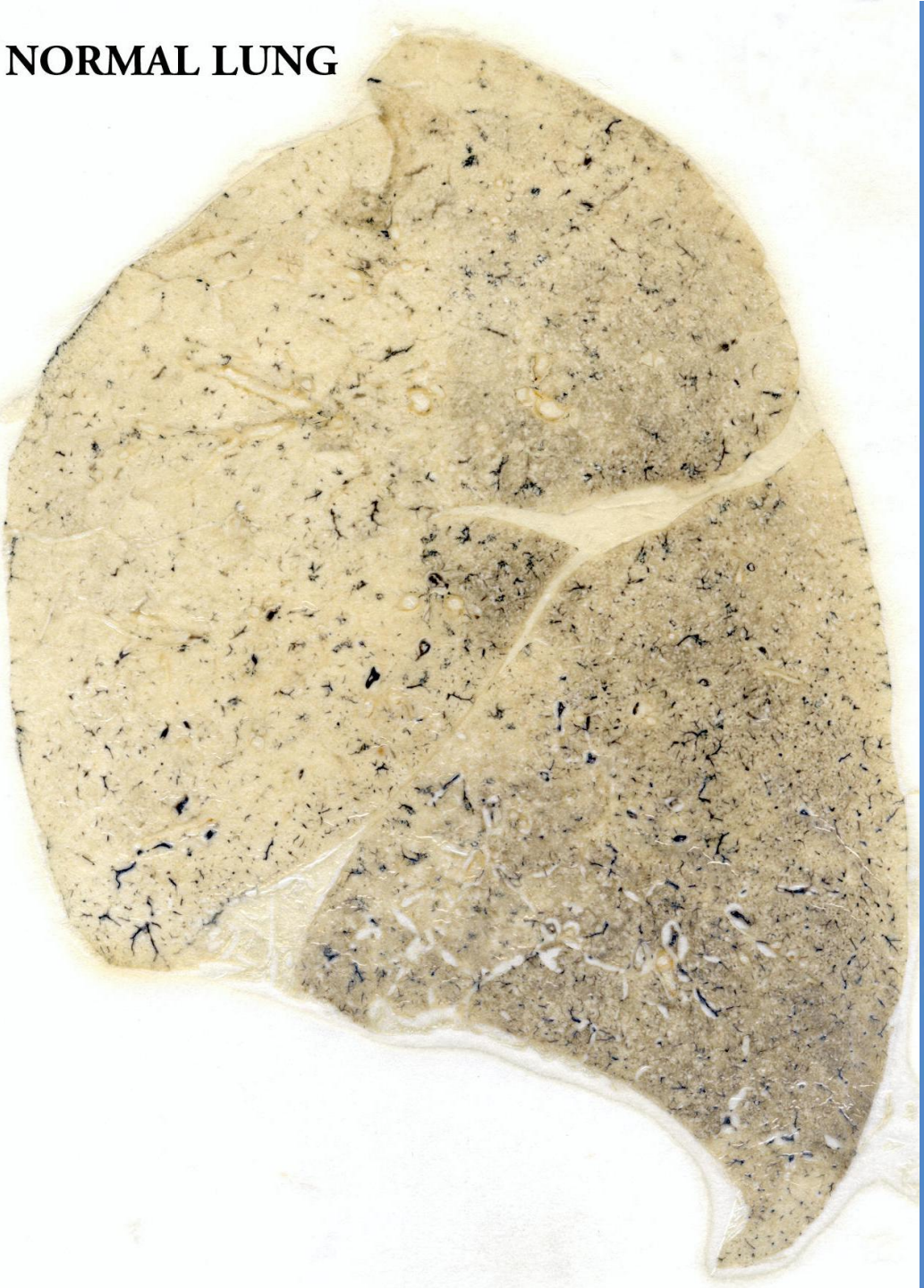


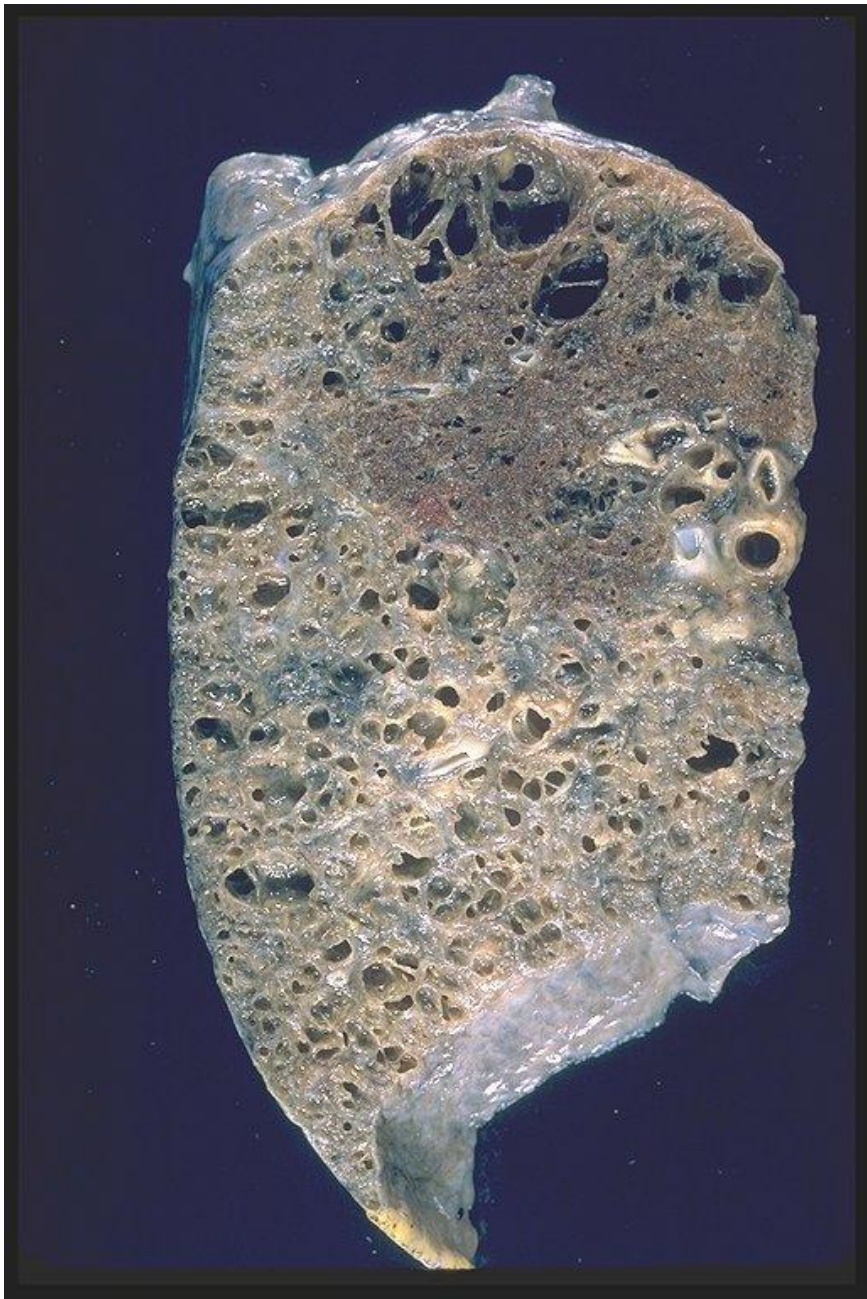




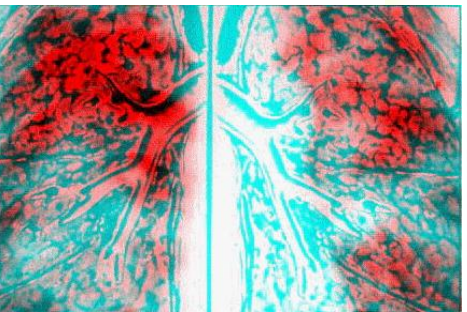
This 50-year-old man presented with end-stage lung fibrosis
PA chest radiograph shows medium to coarse reticular
B: CT scan shows multiple small cysts (honeycombing) involving predominantly the subpleural peripheral regions of lung. Traction bronchiectasis, another sign of end-stage lung fibrosis.

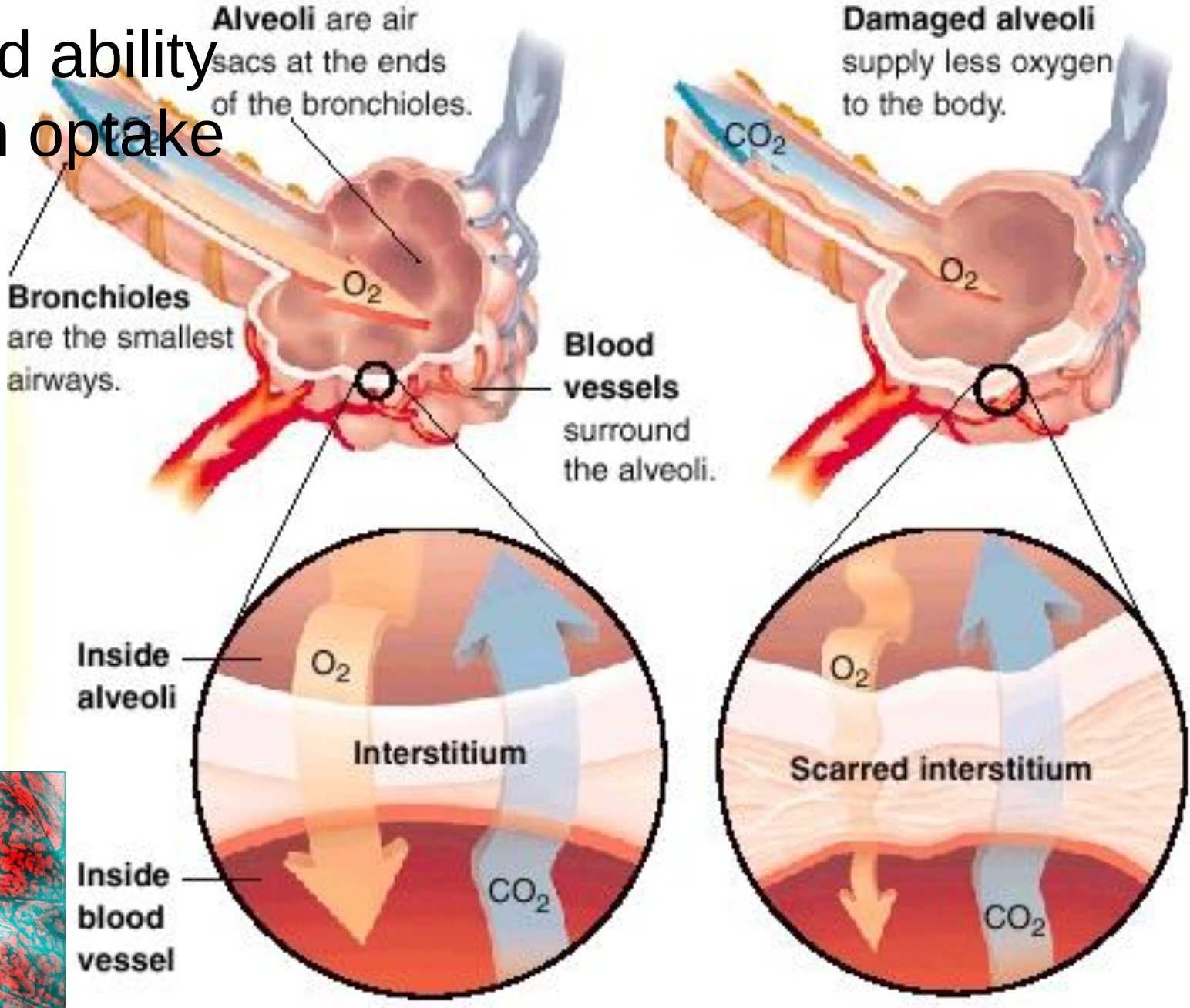
NORMAL LUNG





Lung fibrosis decreased ability of oxygen optake

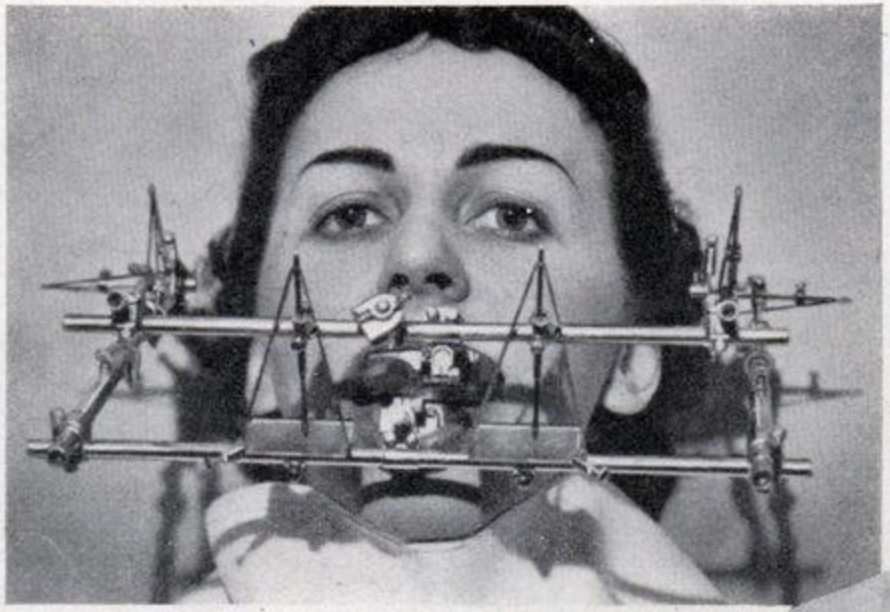




Findings

- Decrease in saturation under activity
  - Later also at rest
- A-gas shows hypoxia not hypercapnia
  - Only in the Terminal fase is hypercapnia seen.
- So no problem giving oxygen (in contrast to COPD)
  - No tendency to develop hypercapnia

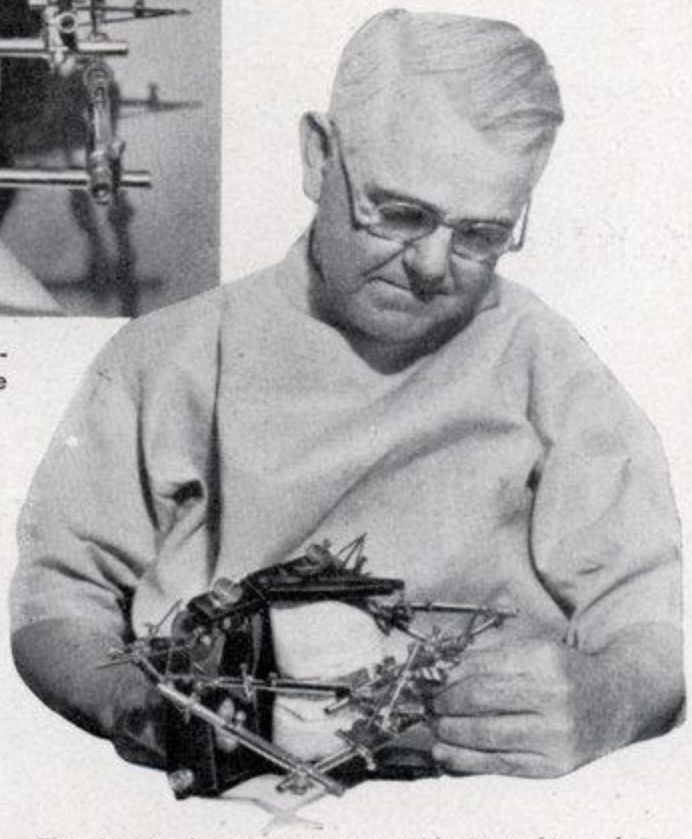
What do we have to measure in patients with lung fibosis??



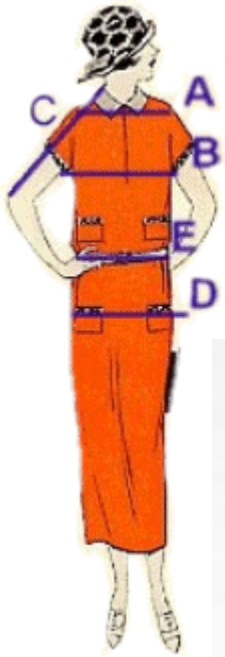
Fitted to a patient's jaws, the "gnathograph" registers the arrangement of teeth and direction of bite

Device Takes Measure of the Teeth

WITH the aid of the "gnathograph," an instrument as mouth-filling as its name, a dentist's patients may now be assured of a perfect fit for artificial teeth. Fitted to the jaws as shown above, the new device registers the arrangement of the teeth and the direction of the "bite," to guide the dentist in straightening teeth or fitting inlays, crowns, bridges, and plates. Its inventor, Dr. Beverly B. McCollum of Los Angeles, Calif., demonstrates in the picture at the right how the instrument is then mounted for use in tooling a plate to just the right shape to give the most comfortable fit in the mouth.



The device then serves as a guide in making plates



How To Measure

There is a slight difference in diagnosing and monitoring the disease

- Lung function
  - Forced volumen
  - TLC, RV and DLCO
- Anatomic changes
  - Bronkoscopy
  - HRCT scan
  - X-RTG Thorax
  - Ekko/hjertekat
  - Lungebiopsy
  - Dexa scanning
- Serological changes
  - Blood tests
- Activity
  - 6 min walking test

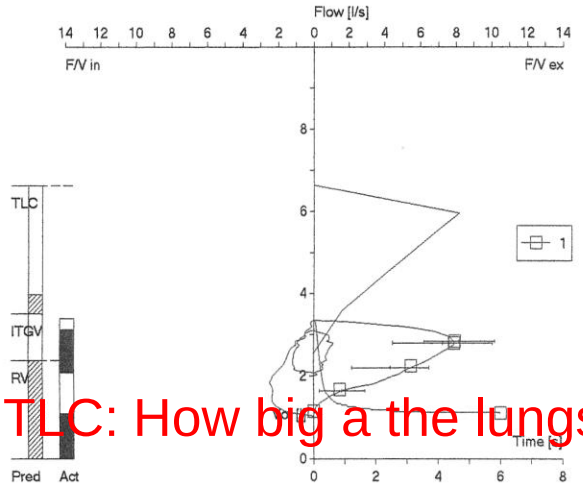
Always initially do TLC;RV and DLCO

Odense Universitetshospital * Medicinsk afd. C * Lungemedicinsk/allergologisk sektion

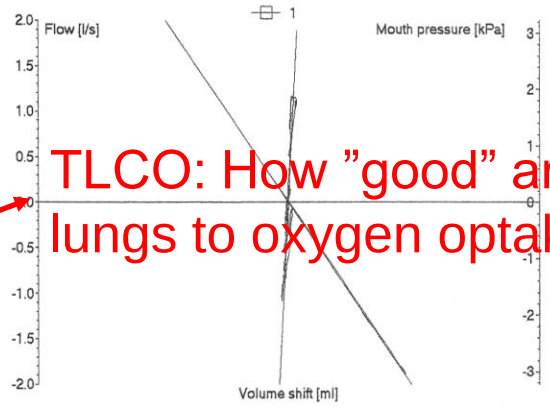
UDVIDET LUNGEFUNKTIONSUNDERSØGELSE

Identification:	120844-
First Name:	Boris Telling
Last Name:	Sørensen
Height:	174,0 cm
Rel. Weight:	92 %
Operator:	
Physician:	--
Last Test:	1

	Pred	Act1	Act2	Act3	(A1/P) %	(Best/Pred)
Date		07-07-2006				
ATS-ok		0.00				
FEV 1	3.22	2.08			64.6	
FVC	4.10	2.24			54.8	
FEV1%F		92.71				
FIV1		1.72				
MIF		0.68				
MEF		0.44				
PIF		2.47				
AIN						
TLC	6.82	3.41			50.0	
RV	2.39	1.09			45.6	
RV%TLC	37.75	31.97			84.7	
Hb		15.40				
TLCOc	9.28	2.97			32.0	
KCOc	1.36	0.98			72.4	



TLC: How big a the lungs ?

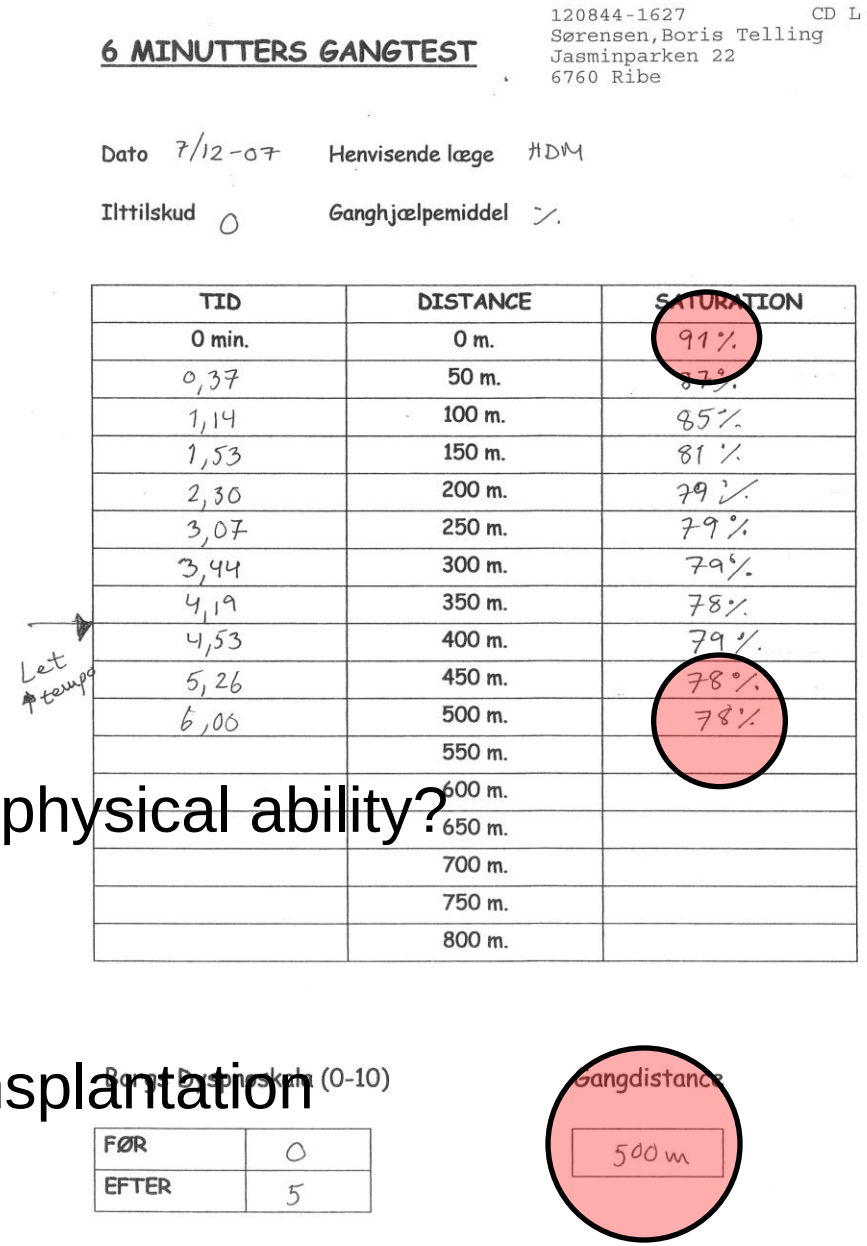


TLCO: How ”good” are the lungs to oxygen optake ?

6-min Walking test

- How far?
- Desaturation?
- Symptoms severe ?

- The test accesses the physical ability?
- Degree of severeness
- Disease development
- Guidance to when transplantation should be considered



6 MINUTTERS GANGTEST

120844-1627 CD L
Sørensen, Boris Telling
Jasminparken 22
6760 Ribe

Dato 7/12-07 Henvisende læge HDM

Ilttilskud 0 Ganghjælpemiddel -

TID	DISTANCE	SATURATION
0 min.	0 m.	91 %
0,37	50 m.	87 %
1,14	100 m.	85 %
1,53	150 m.	81 %
2,30	200 m.	79 %
3,07	250 m.	79 %
3,44	300 m.	79 %
4,19	350 m.	78 %
4,53	400 m.	79 %
5,26	450 m.	78 %
6,00	500 m.	78 %
	550 m.	
	600 m.	
	650 m.	
	700 m.	
	750 m.	
	800 m.	

Let tempo

Borg Dyspnoeskala (0-10)

FØR	0
EFTER	5

Gangdistance
500 m

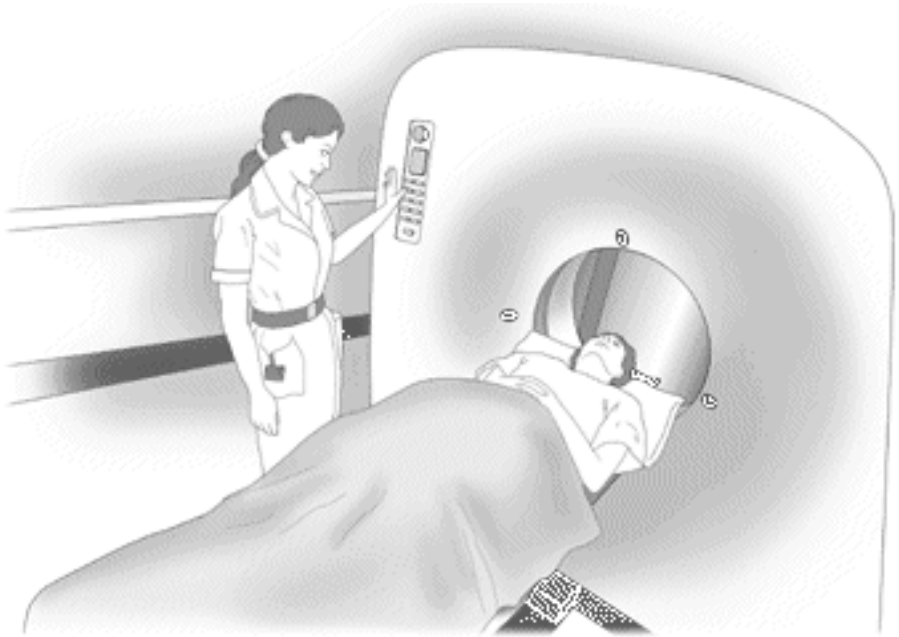
Mette Aukjær

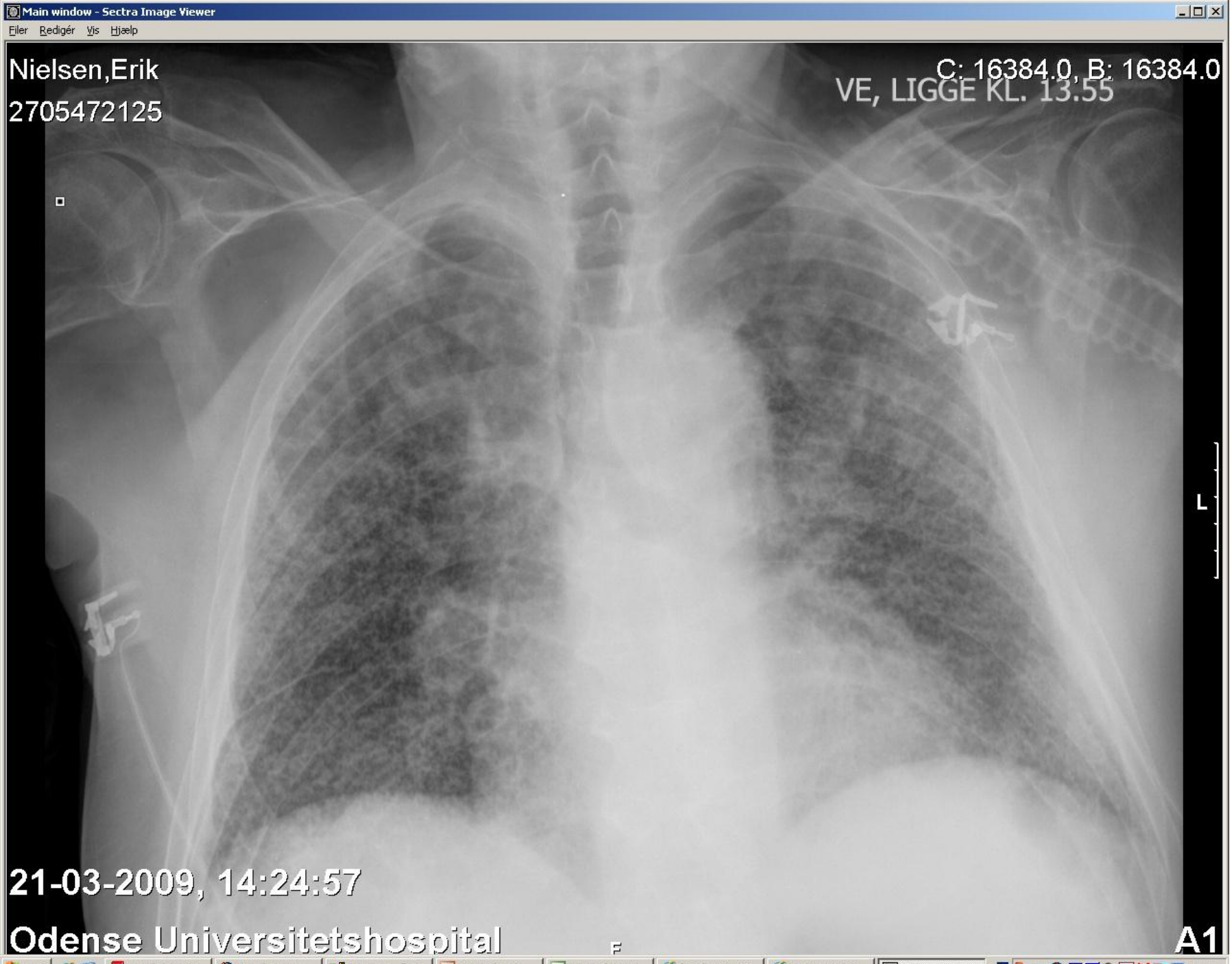
HRCT- scan

- Changes
- distribution
- Pattern

Which disease ?
Who bad ?
Diffential diagnosis ?
Further work-up ?

- development
  - Effect of treatment
  - Changes in disease

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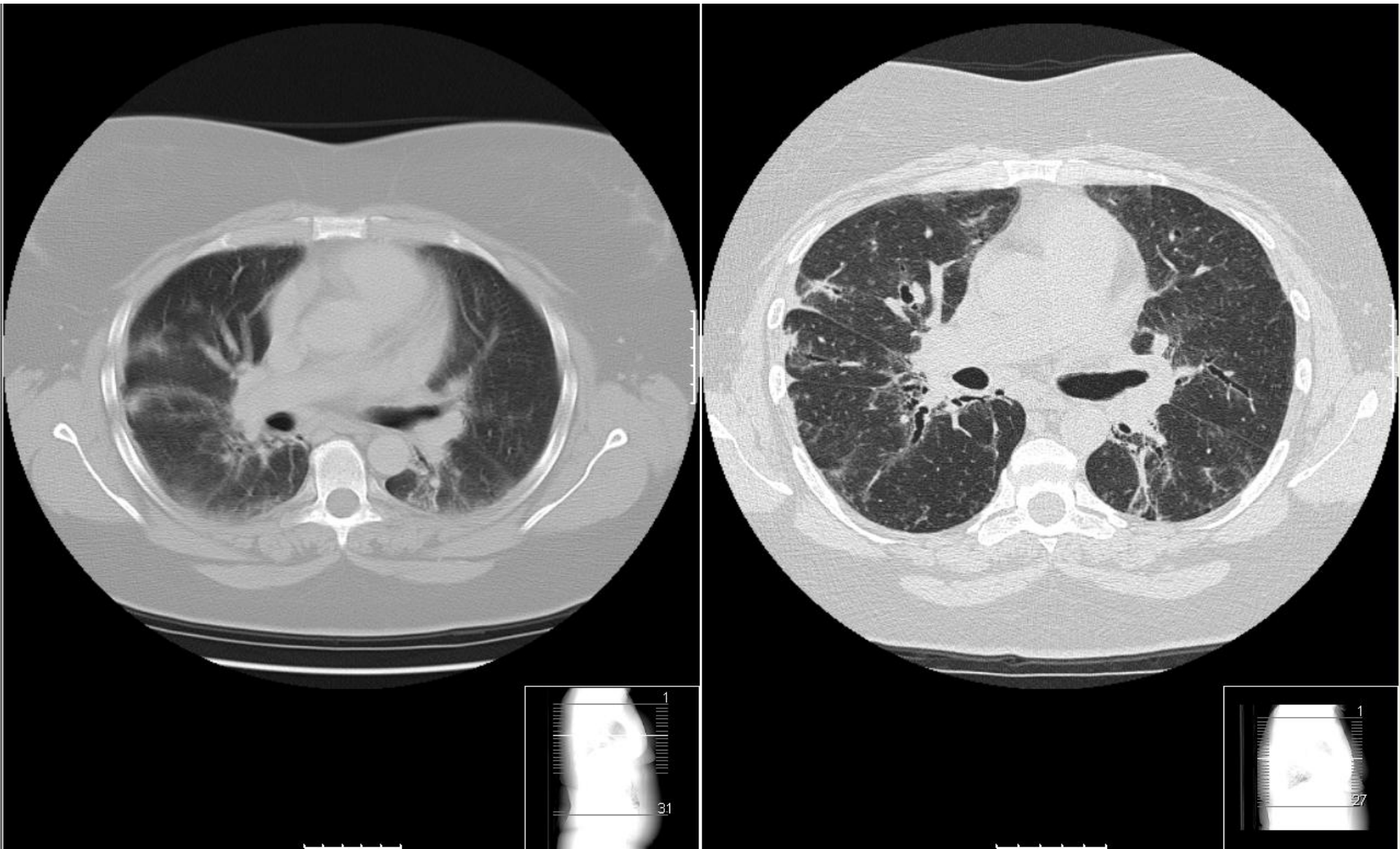
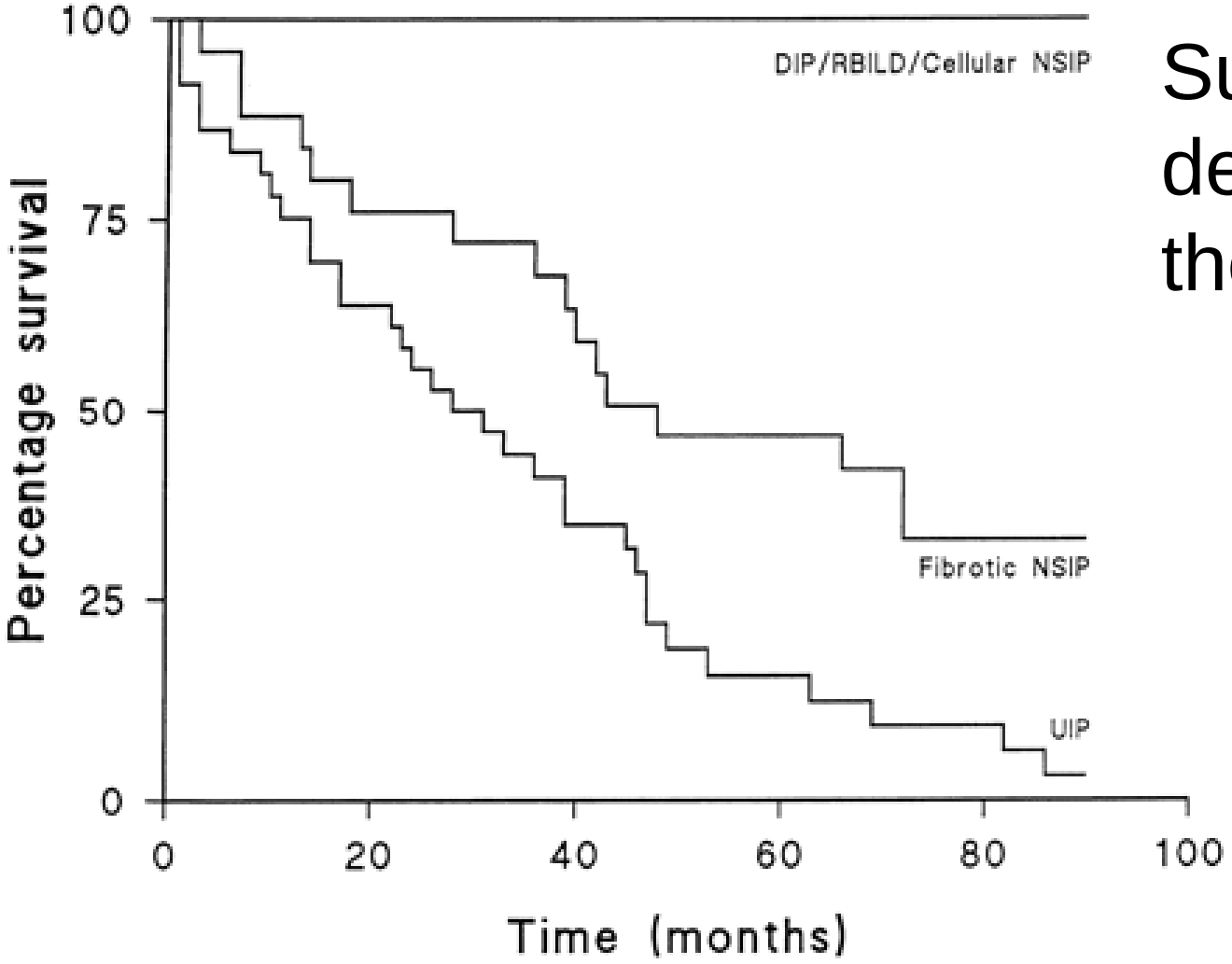
CT versus HRCT

Always HRCT !!!





Survival is dependent on the diagnosis

usual interstitial pneumonia (UIP), fibrotic nonspecific interstitial pneumonia (NSIP), and desquamative interstitial pneumonia (DIP)/respiratory bronchiolitis-associated interstitial lung disease (RBILD)/cellular NSIP

Am. J. Respir. Crit. Care Med., Volume 162, Number 6, Dec. 2000, 2213-2217

Restrictive diseases

Intrinsic lung diseases

- Interstitial lung diseases
  - » Arthritis related (SLE, RA, scleroderma)
  - » "Ideopatic" (ex UIP)
    - "smoke related" (ex Histeocytosis X)
- Asbestosis/silicosis
- Allergic (allergic alveolitis)
- Pleura (debris-exsudat)
- Medicine (nitrofurantoin, amiodarone, bleomycin).
- Pneumonia
- radiation

Extrinsic diseases (extra-parenchymale diseases)

- Non-neuromuskular
  - Deformities
  - Heart disease
  - ARDS
- Neuromuscular
  - Poliomyelitis, Guillain-Barre syndrome, ALS, myasthenia gravis, muscular dystrophies

Inflammation and/or scarring of lung tissue

reduced space or muscular power

Fill airspaces exudat/debris (pneumonnitis)

Treatment

- Immunosupresiva
  - Perferidine
  - Prednisolon
    - One time
    - Continuos
  - Others
    - Azatioprime; metrotrexate, cyclosporine many others
- Anti-inflammatory
  - acetylcysteine
- Removal of cause
  - Allergic alveolitis
  - Langerhans histeocytosis X;REBILD


LAST
CHANCE

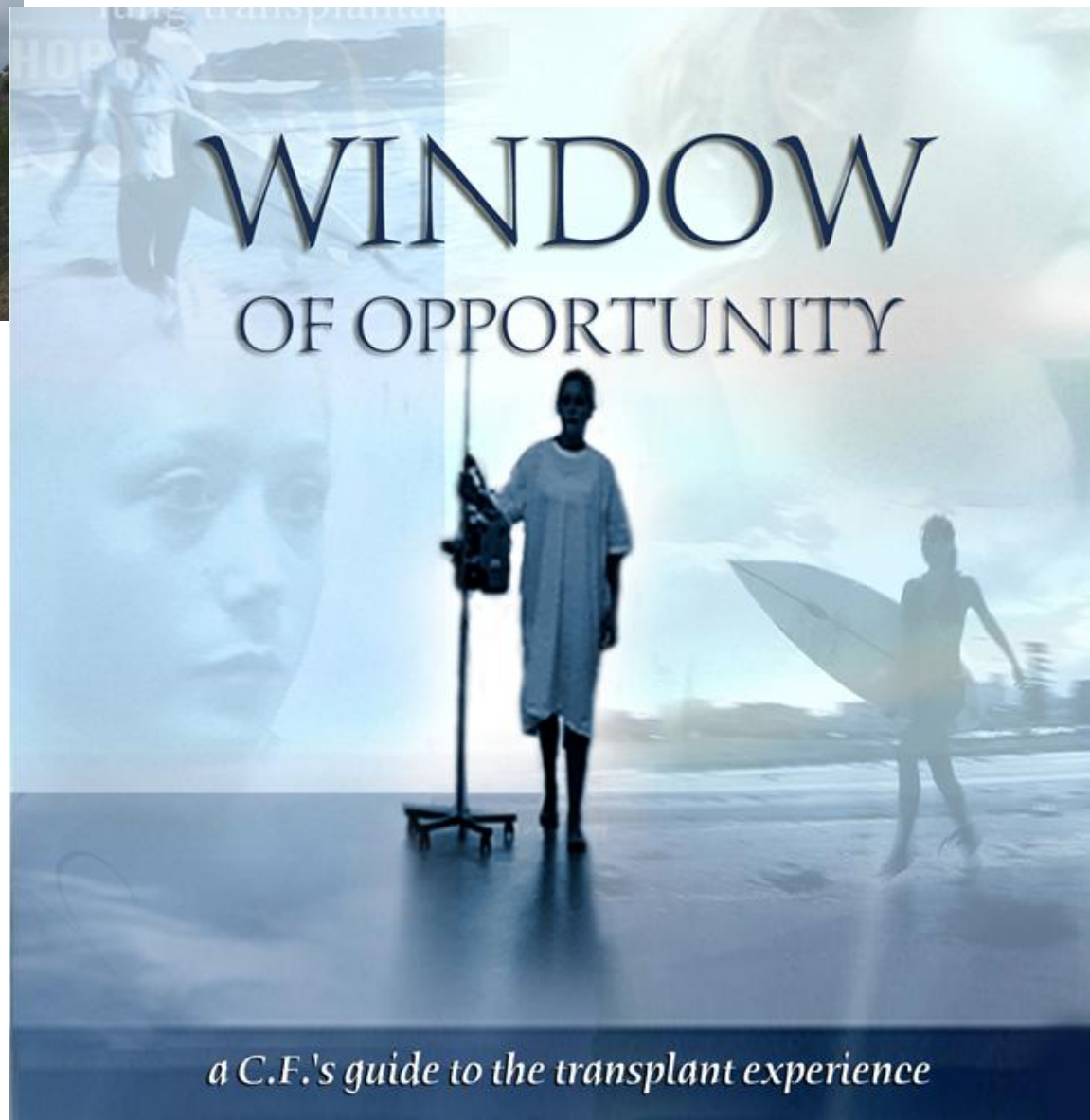

WINDOW
OF OPPORTUNITY
a C.F.'s guide to the transplant experience

TABLE 1. DISEASE-SPECIFIC GUIDELINES FOR REFERRAL FOR LUNG TRANSPLANTATION.*

Chronic obstructive pulmonary disease

FEV_1 <25 percent of predicted value after bronchodilator therapy
Clinically significant hypoxemia, hypercapnia, or pulmonary hypertension; rapid decline in lung function; or frequent severe exacerbations

Idiopathic pulmonary fibrosis

Symptomatic disease unresponsive to medical therapy
Vital capacity <60 to 70 percent of predicted value
Evidence of resting or exercise-induced hypoxemia

Cystic fibrosis

FEV_1 ≤30 percent of predicted value
FEV_1 >30 percent with rapidly declining lung function, frequent severe exacerbations, or progressive weight loss
Female sex and age of less than 18 years with FEV_1 >30 percent†

Primary pulmonary hypertension

NYHA functional class III or IV
Mean pulmonary-artery pressure >55 mm Hg
Mean right atrial pressure >15 mm Hg
Cardiac index <2 liters/min/m^2
Failure of medical therapy, especially intravenous epoprostenol, to improve NYHA functional class or hemodynamic indexes

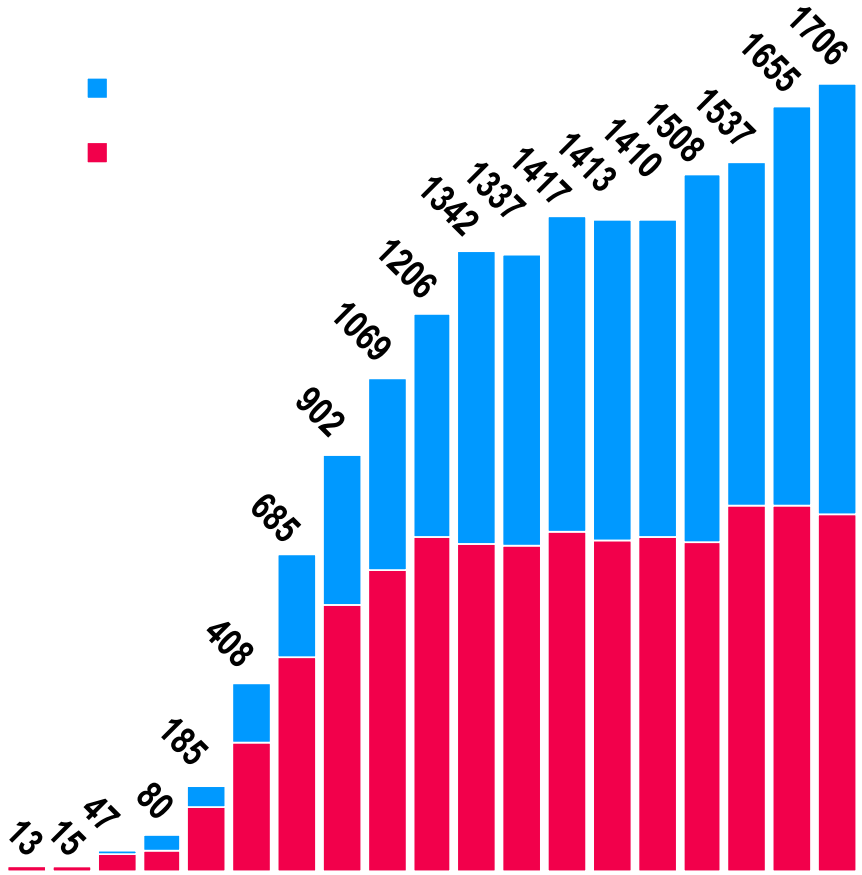
Eisenmenger's syndrome

NYHA functional class III or IV despite optimal medical management

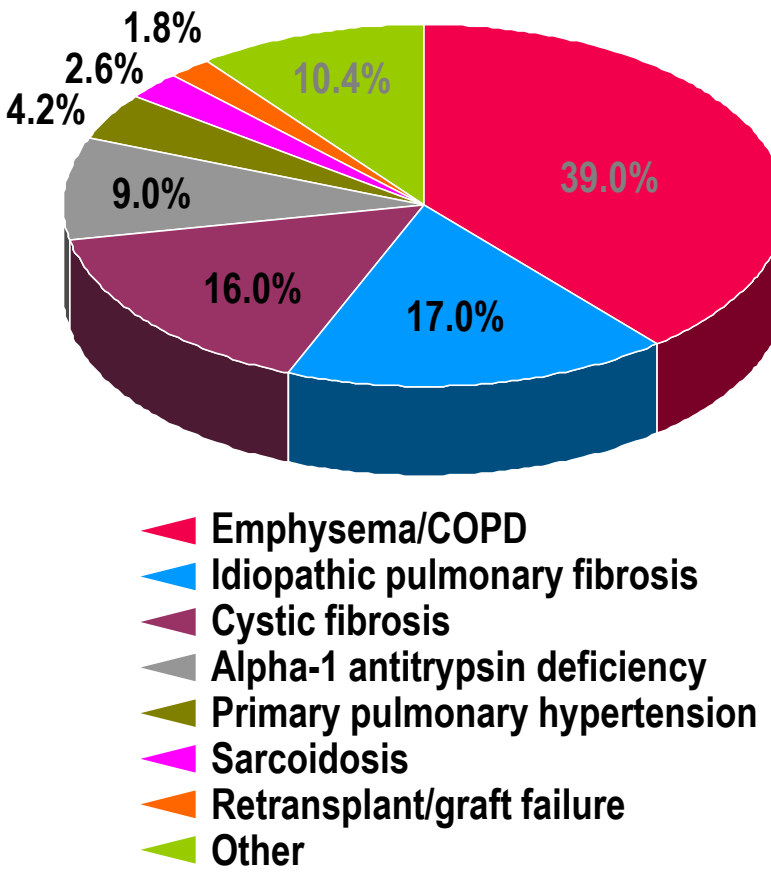
- transplantation is dependent on the disease

Worldwide Lung Transplantation Numbers

Lung transplants performed worldwide, by year



Primary diagnosis, 01/1995 - 06/2003



Source: International Society of Heart and Lung Transplantation (ISHLT); UNOS

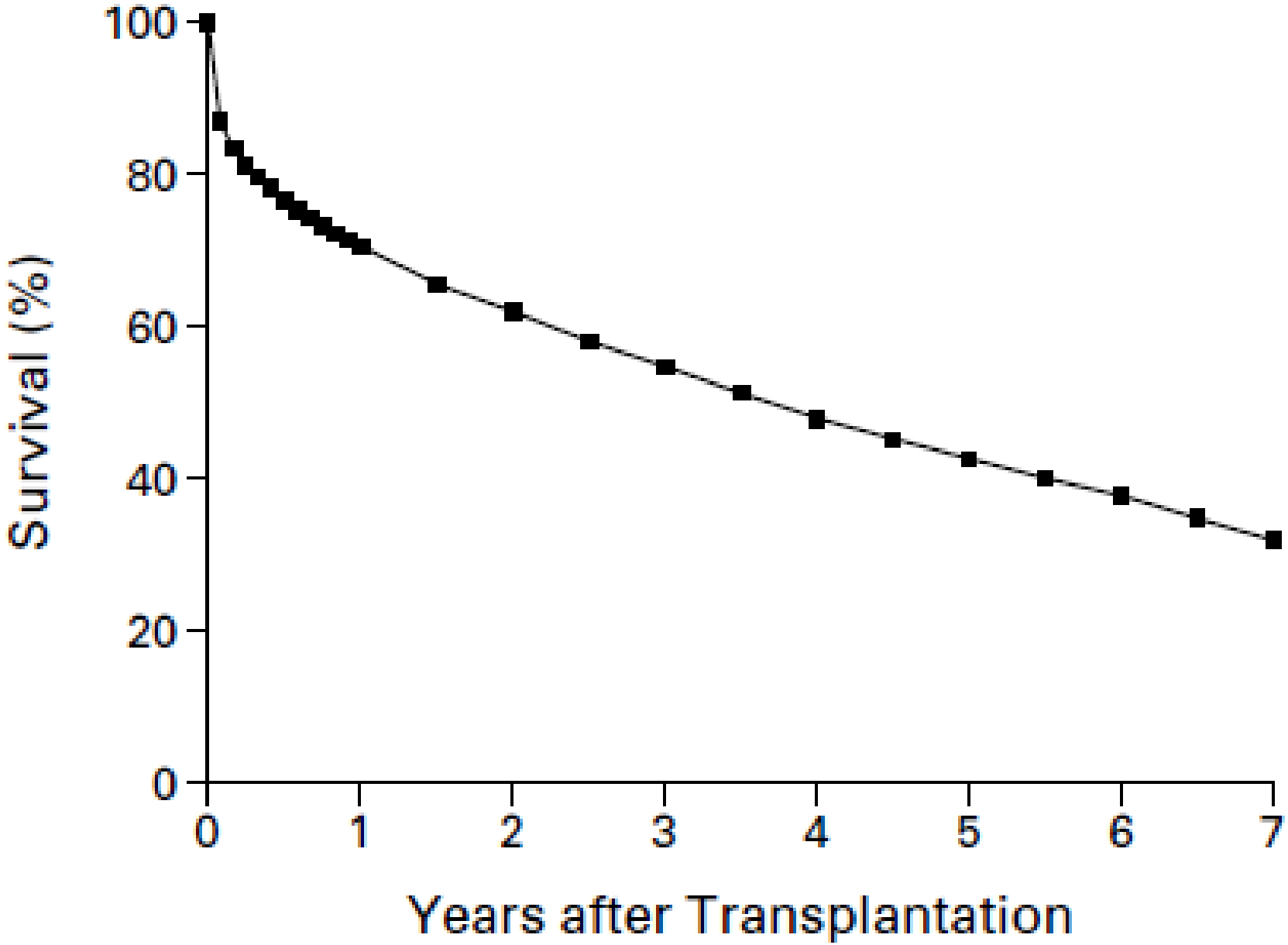
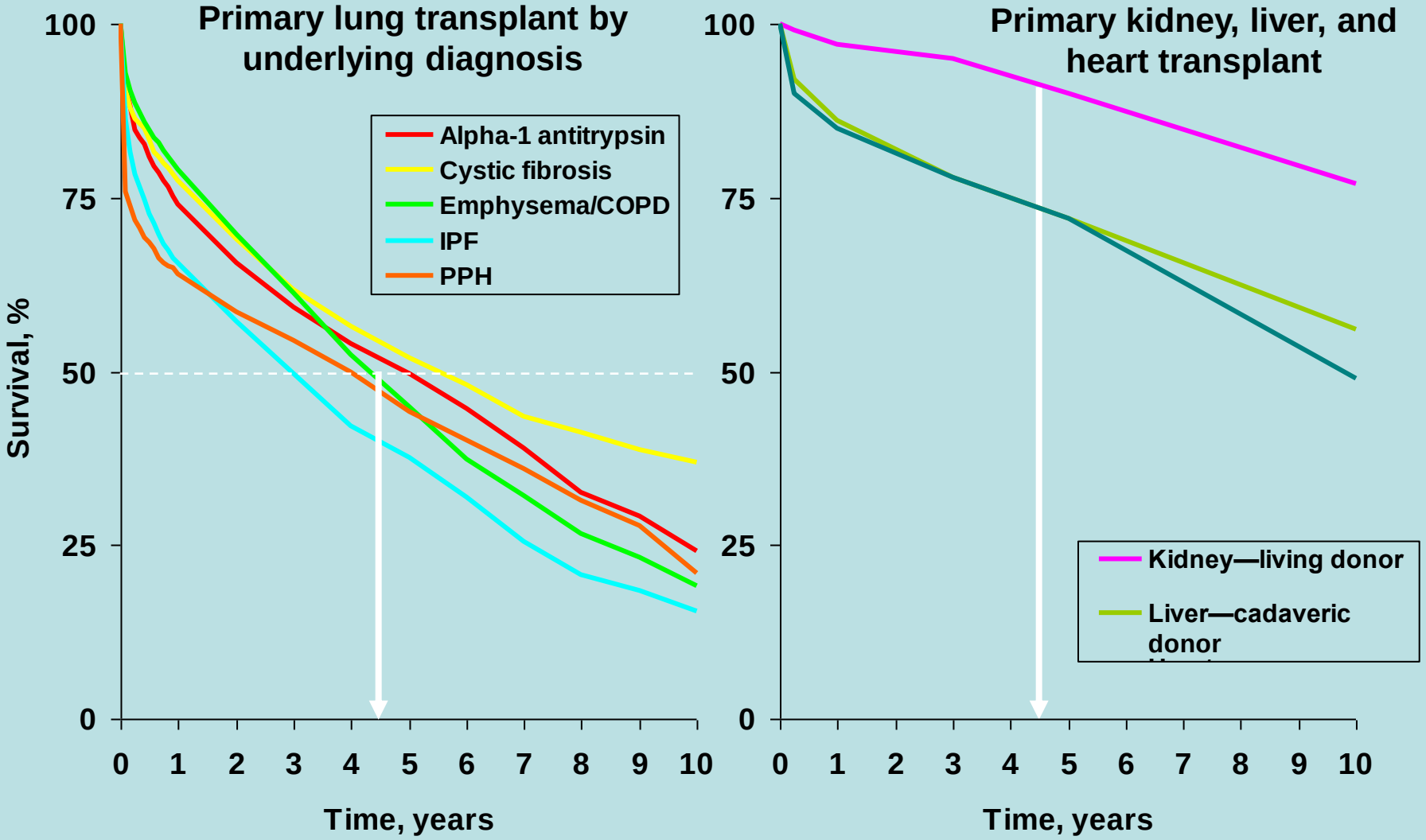


Figure 2. Actuarial Survival after Lung Transplantation.

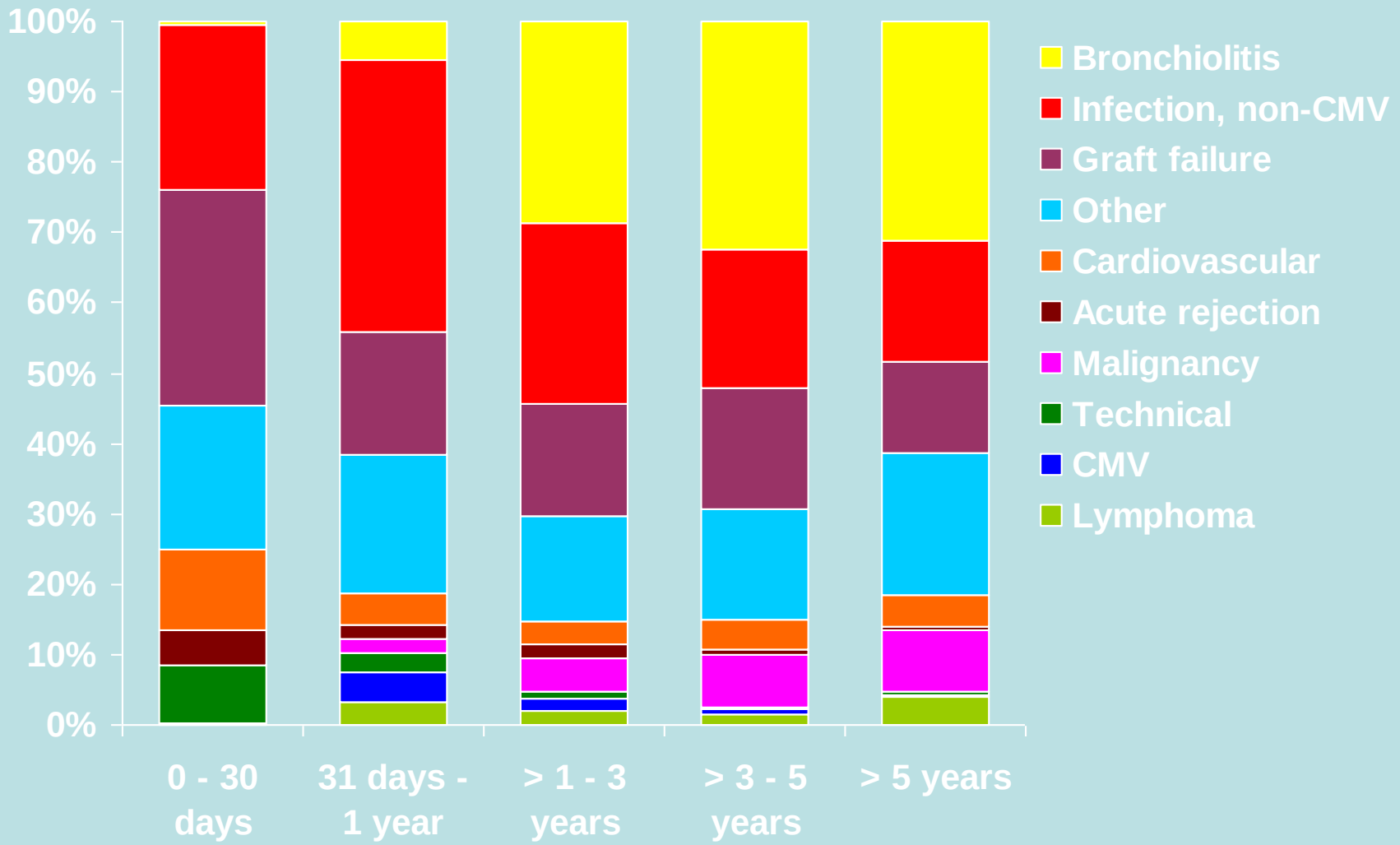
CM-106

Comparative Transplantation Survival Rates



*Kidney, liver, and heart data extrapolated from OPTN Annual Report, 2003.

Causes of Death Following Lung Transplantation



Status for Lung transplantation

- Survivel —50% died after 5 years
- Bronchiolitis obliterans main reason for a bad survivel rate
- Main aim to treat and prevent bronchiolitis obliterans

More time???