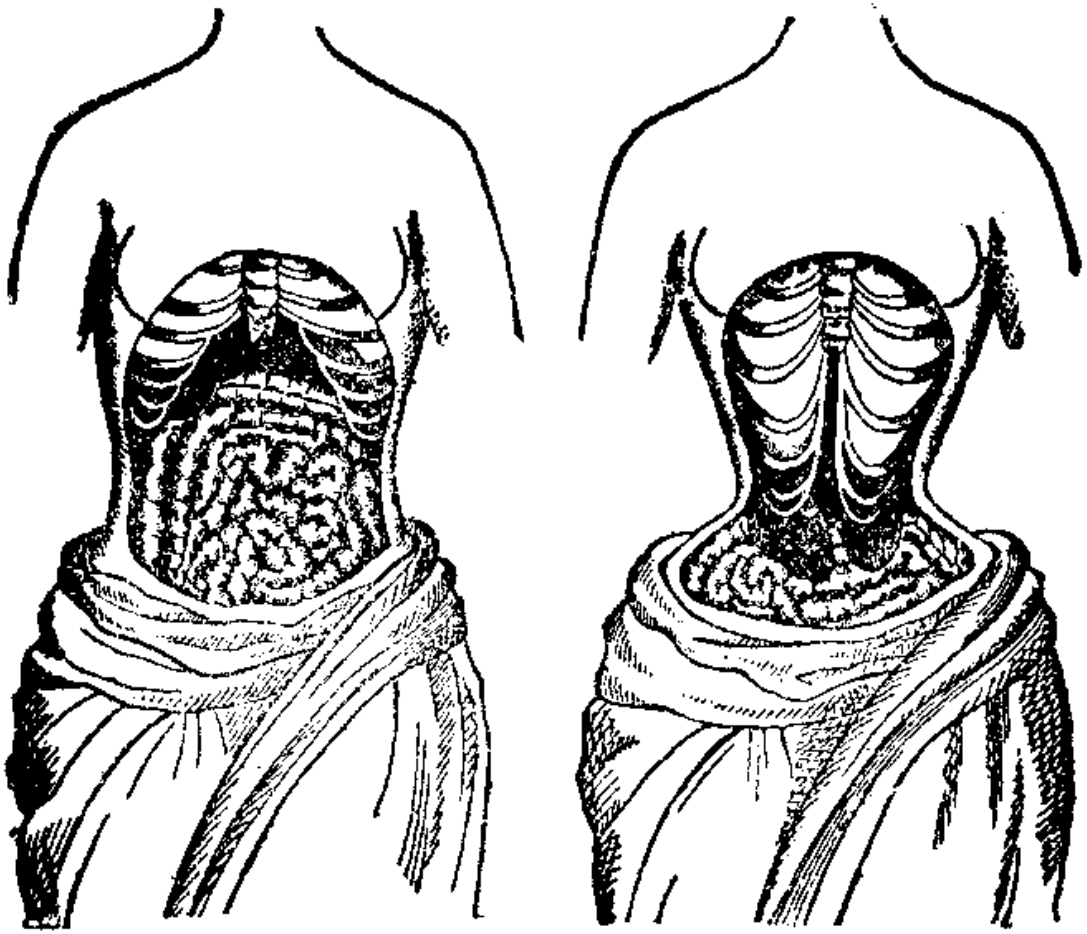
Interstitial lung diseases
Restrictive lung diseases
Lung fibrosis

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

”small” lungs

Finn Rasmussen

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 a ”acute” restriktive disorder

Pleuraexsudate, pneumonia
Atelektasis etc..

However. Normally in lungfibrosis

Disease localized in the paremkyma

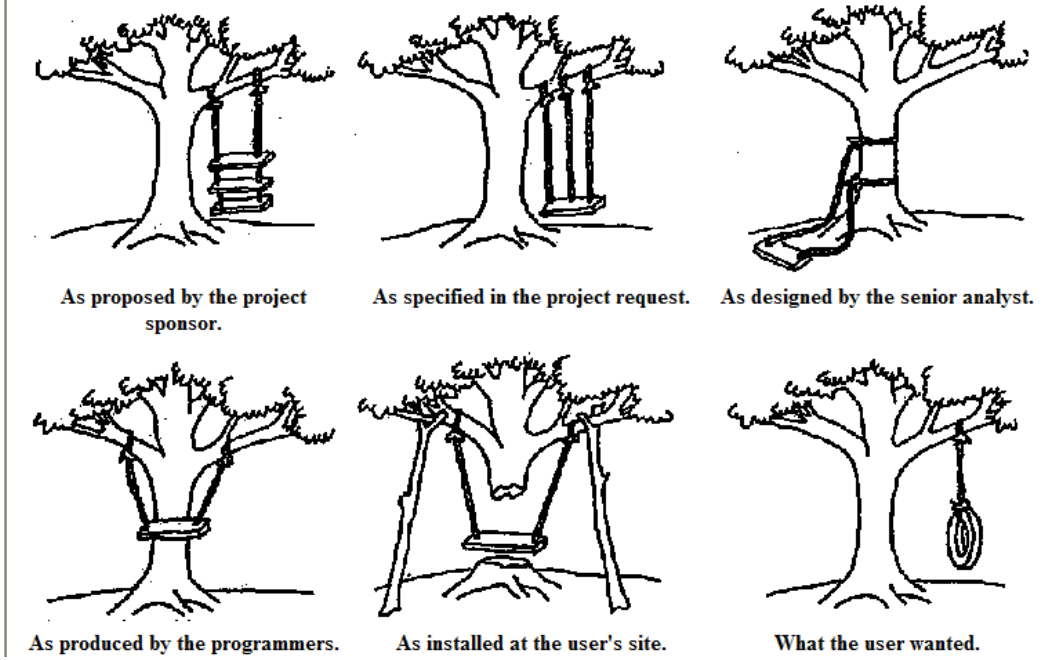
What is lost is forever lost …..

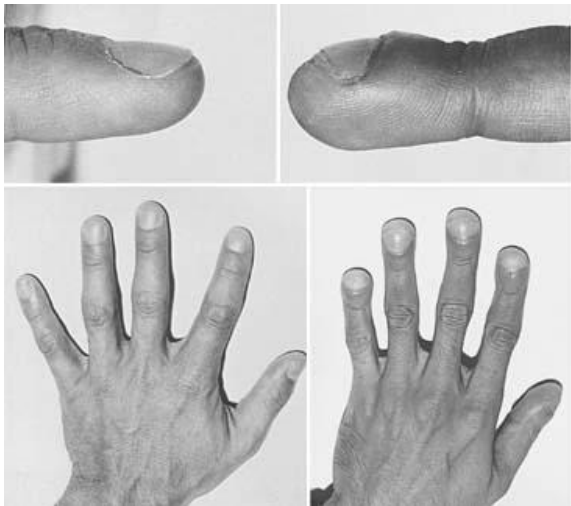
…if treatment is not started in time…

Symptoms

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

- Signs
  - cyanosis
  - Low saturation
- low lung function
- Dromstikfingers
- Velcro sound at stethoscopy



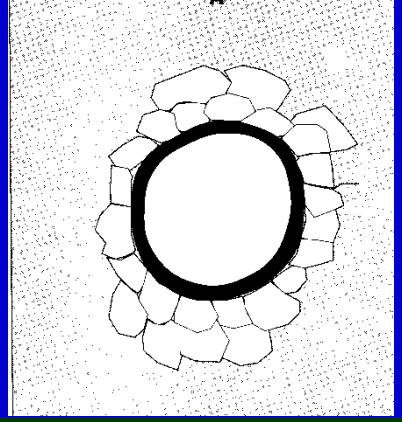


D'oh!

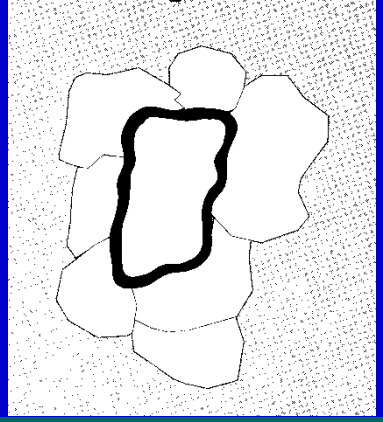
However the initial most often mistake diagnosis that mis-interpretated as lung fibrosis is ???

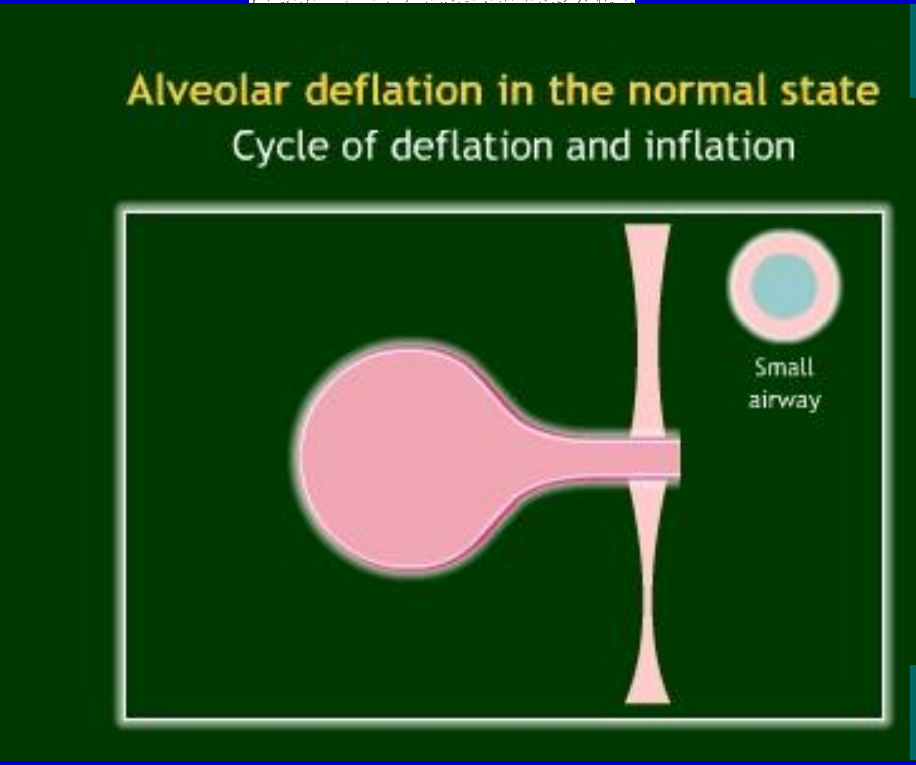
COPD !!!!

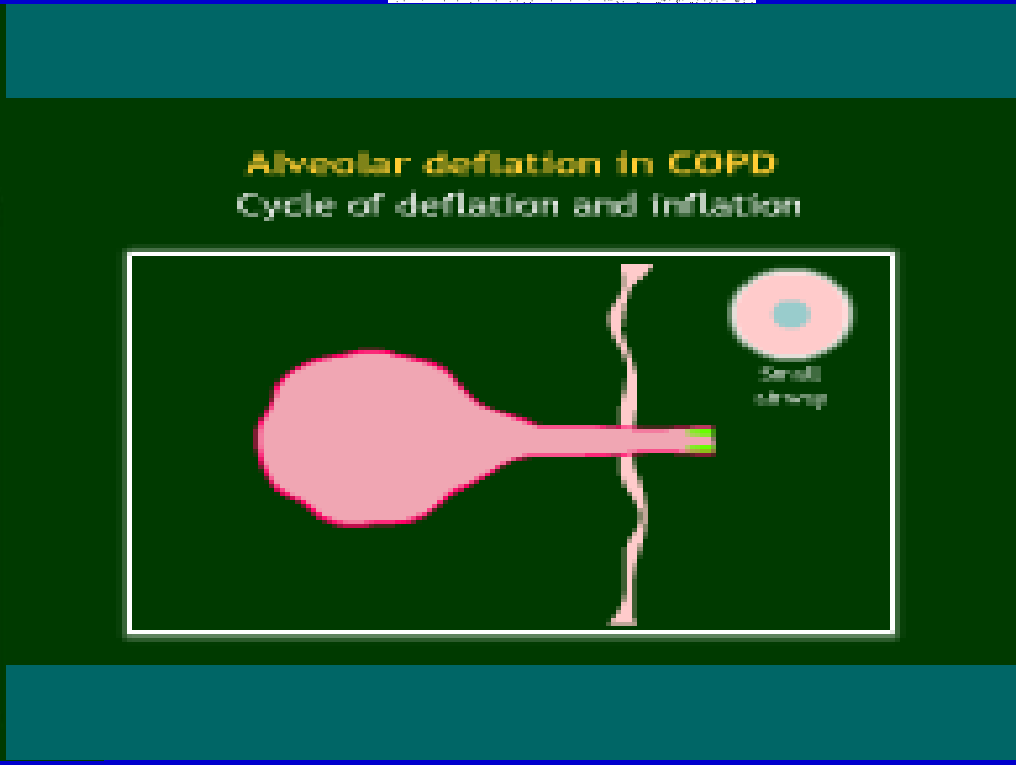
Why??



COPD
FVC is falsely decreased







"Air trapping"
"solved" by Slow vital capasity
FVC<SVC
Eller TLC

Odense Universitetshospital

Lungemedicinsk amb.: Lungefunktionsundersøgelse

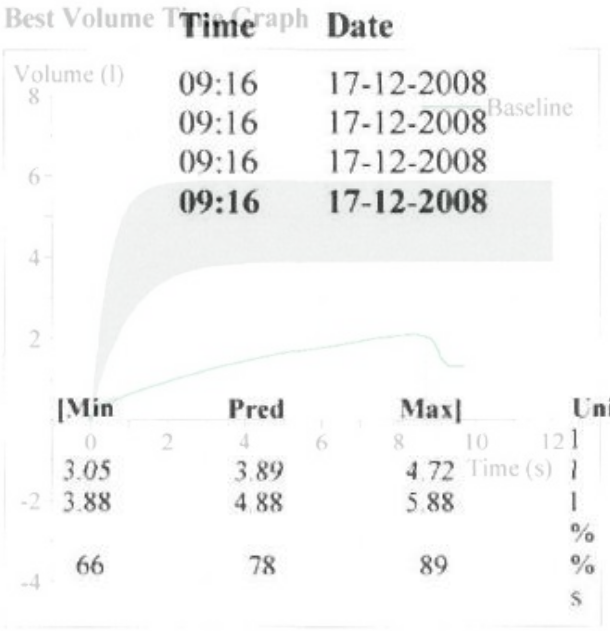
17-12-2008

ID:	060955-1461	**Alias ID:**	
Name:	**Vadsř-Jensen, Knud Erik**		
Age:	53 (06-09-1955)	**Exam Date:**	17-12-2008 09:16
Gender:	Male	**Origin:**	Caucasian **Factor:** 100
Height:	184 cm.	**Weight:**	57.0 kg. **Smoker:** 0
Referred by:		**Occupation:**	

	VC	FEV1	FVC	PEF	VAR	Quality	Time	Date
Base		0.57	1.78	157	-14 %	Good Blow	09:16	17-12-2008
Base		0.61	2.12	168	+0 %	Good Blow	09:16	17-12-2008
Base		0.57	1.79	169	-14 %	Good Blow	09:16	17-12-2008
+ Base		**0.61**	**2.12**	**169**	**+0 %**		**09:16**	**17-12-2008**

ATS/ERS Criteria (2005): Tests do not meet repeatability standards for FVC.

Variation is based on FEV1 + FVC



Index	Base	%Pred	Post 1	%Pred	Change	[Min	Pred	Max]	Units
VC						3.05	3.89	4.72	l
FEV1	0.61	16				3.88	4.88	5.88	l
FVC	2.12	43							%
FEV1/VC						66	78	89	%
FEV1/FVC	29								s
FET	8.70								

One clue is that the pt is obstruktive

Then wounder !!

Sa02 92

puls 90-120

Udvidet Lungefunktionsundersøgelse

dentifikation:	060955-1461	Fornavn:	Knud Erik	Efternavn:	Vadsø-Jensen
Fødselsdato:	06-09-1955	Alder:	53 Years	Køn:	male
Højde:	184,0 cm	Vægt:	60,0 kg	Referencer:	Standard

Spirometri:

		Test 1	% af forv.	Test 2	% af forv.	LL	Forventet	UL
Dato		05-01-09						
VC MAX	[L]	2.30	45.1 %			4.17	5.09	6.01
FVC	[L]	1.77	36.3 %			3.88	4.88	5.88
FEV 1	[L]	0.80	20.5 %			3.05	3.88	4.72
FEV 1 % FVC	[%]	44.98						
FEV 1 % VC MAX	[%]	34.73	44.7 %			65.91	77.67	89.43
PEF	[L/s]	3.11	34.0 %			7.18	9.17	11.15

Kropspletysmografi:

		Test 1	% af forv.	Test 2	% af forv.	LL	Forventet	UL
TLC	[L]	8.78	115.1 %			6.47	7.62	8.77
RV	[L]	6.26	266.9 %			1.67	2.35	3.02

TLC is normal

Diffusionsbestemmelse:

		Test 1	% af forv.	Test 2	% af forv.	LL	Forventet	UL
TLCO SB	[mmol/min/kPa]	1.12	10.3 %			8.60	10.91	13.23
TLCOc SB	[mmol/min/kPa]	1.14	10.5 %			8.60	10.91	13.23
VA	[L]	3.83	51.3 %			7.47	7.47	7.47
TLCO/VA	[mmol/min/kPa/L]	0.29	20.5 %			1.43	1.43	1.43
TLCOc/VA	[mmol/min/kPa/L]	0.30	20.8 %			1.43	1.43	1.43
Hb	[mmol/L]	8.70						

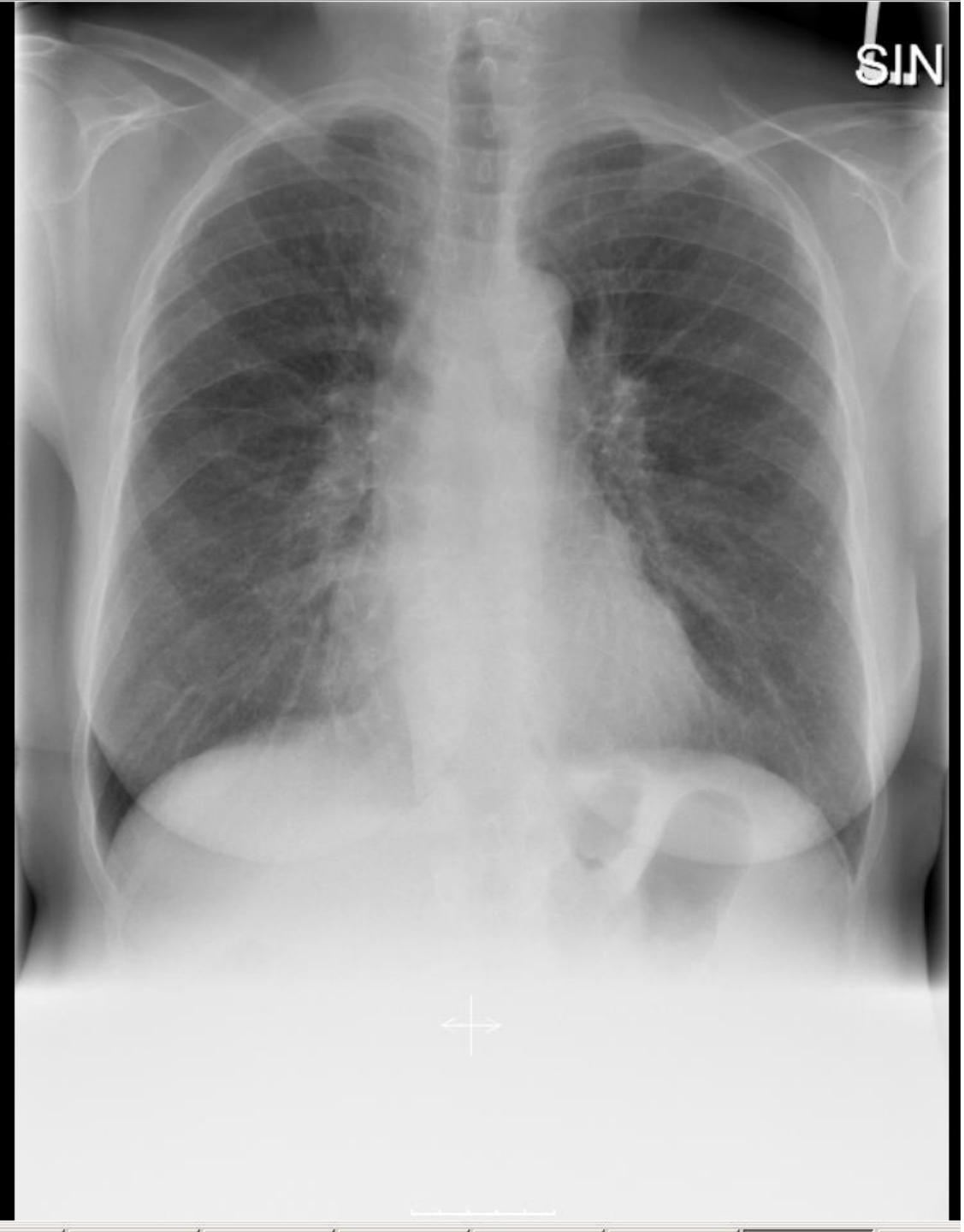
Myths !

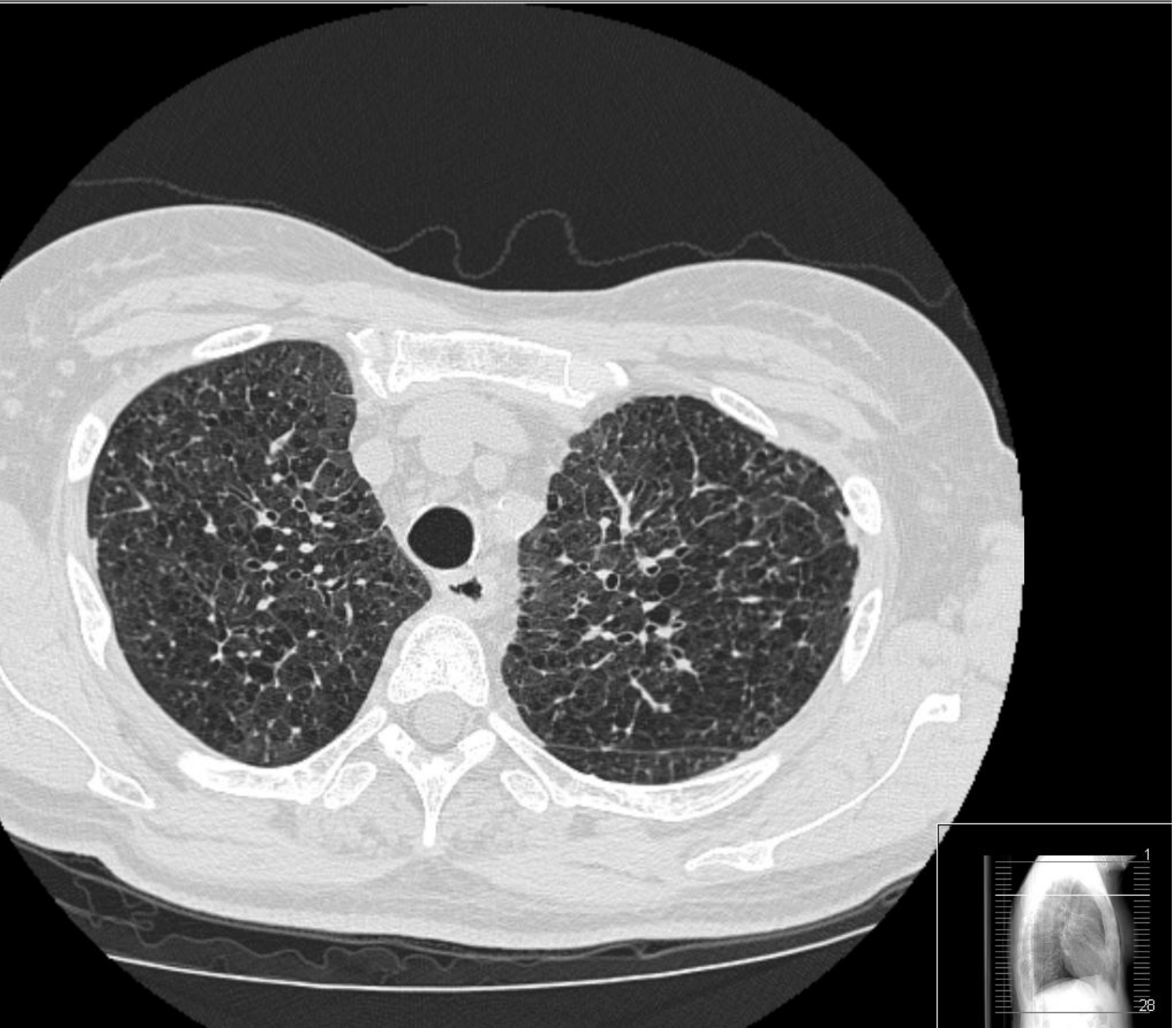
- Smoking does NOT normally cause lung fibrosis
- However nothing without exceptions ..... Very rare lung diseases ……
- Smoking gives lungfibrosis…..???!!!

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


SIN


1
28


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

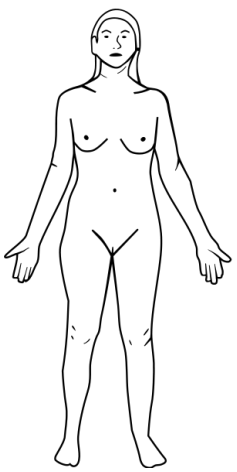
Causes:
Restictive lung diseases

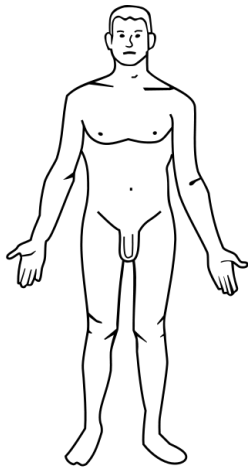
1. Changes in the lung parenkyma

=Lung fibrosis

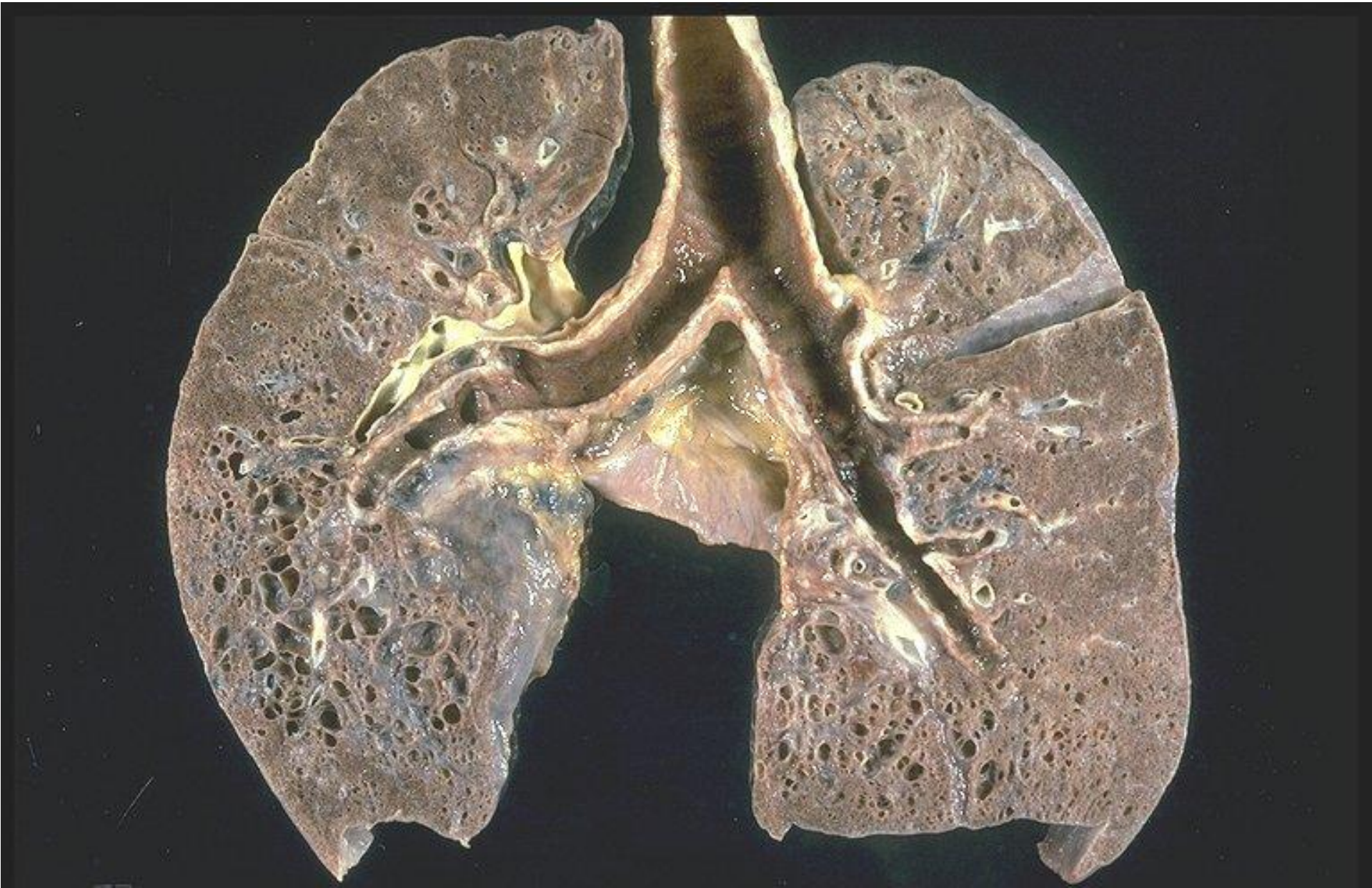
2. Diseases in the pleura, chest wall, muscles and nerves

What happens in lung fibrosis ??

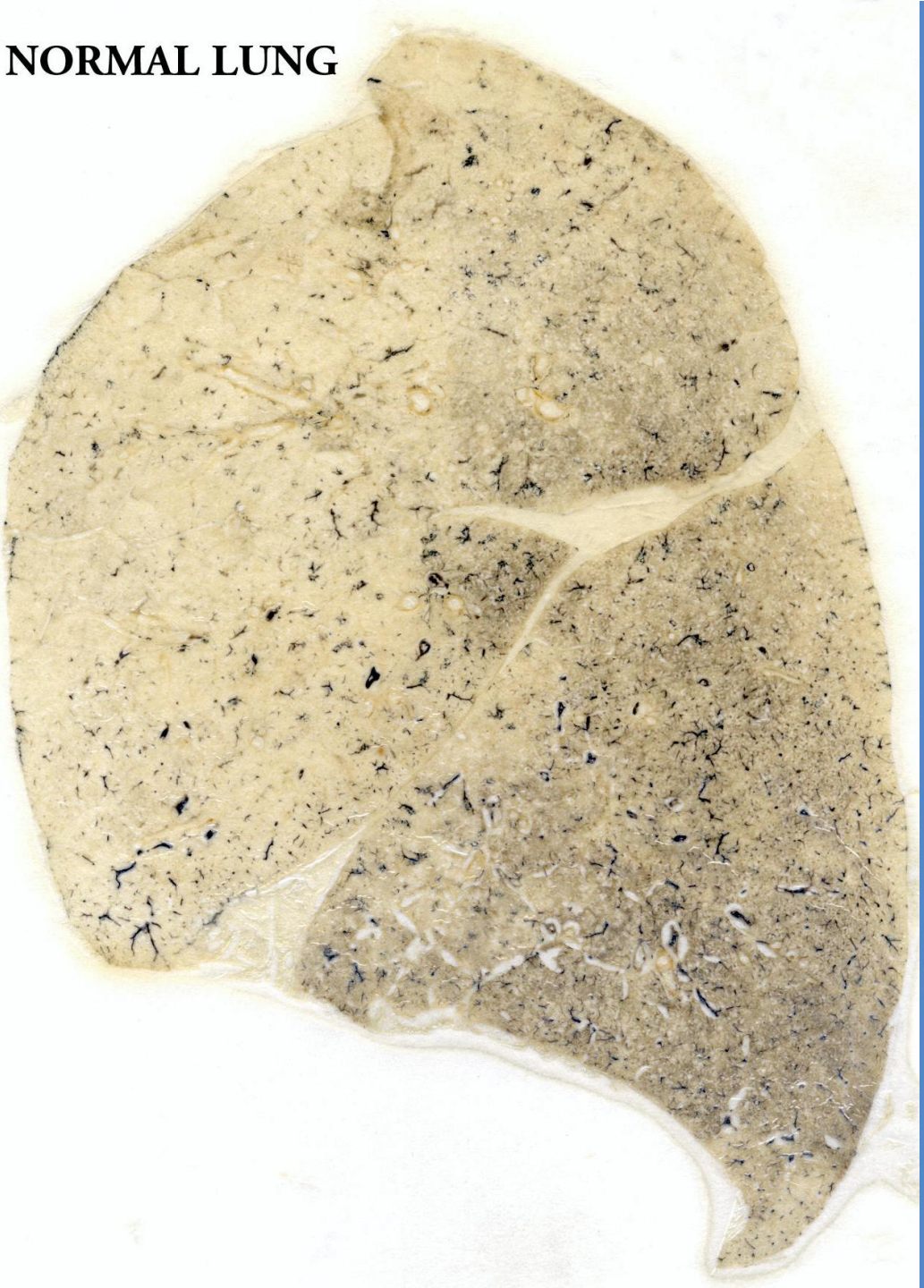




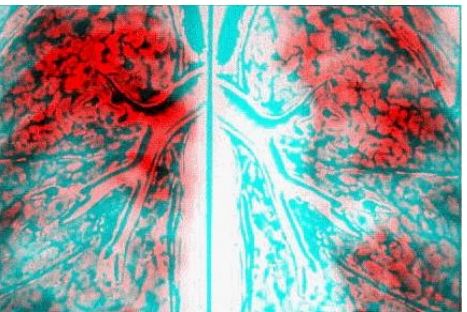
Scar tissue (fibrosis) in the lungs

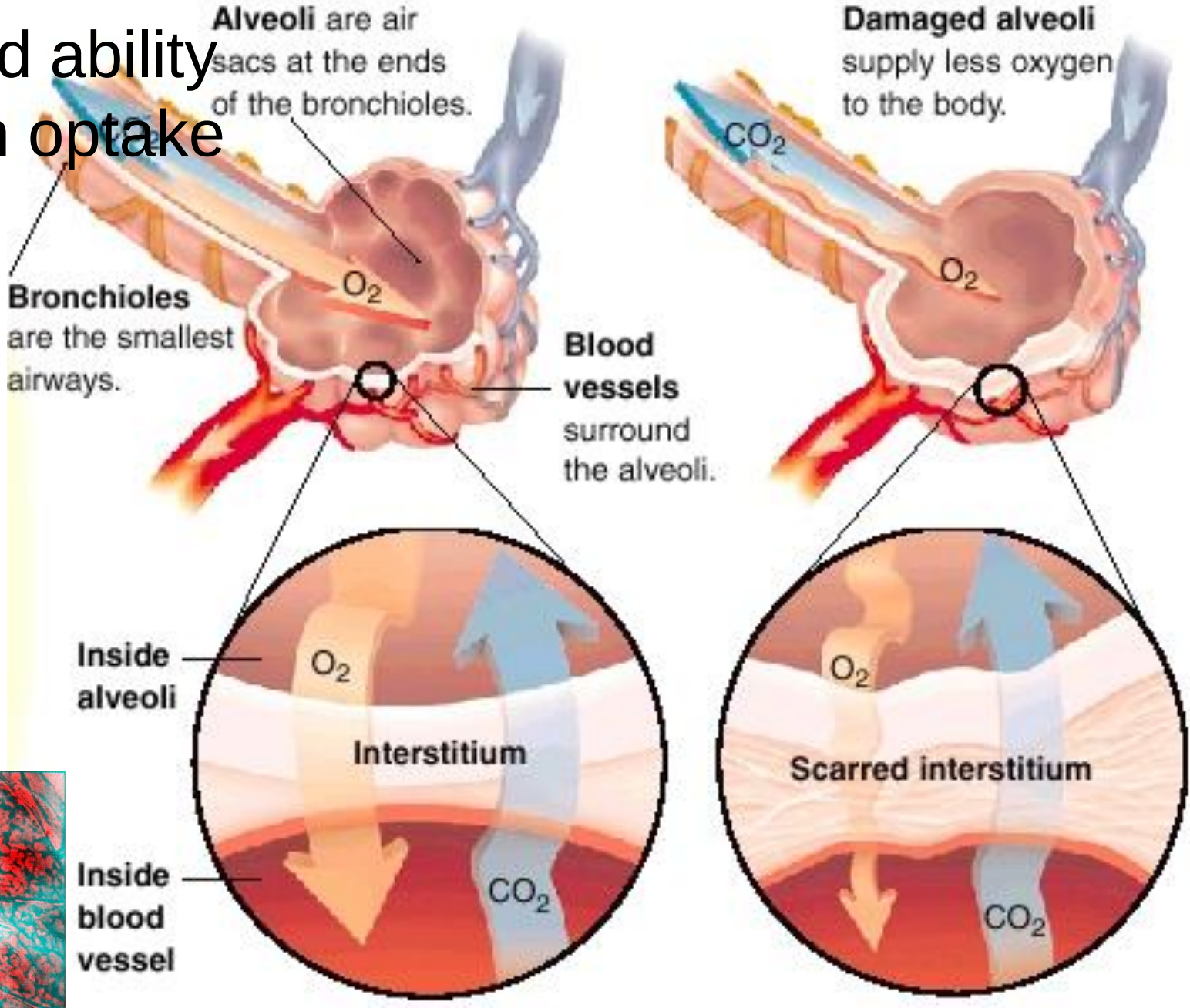


NORMAL LUNG



Lung fibrosis decreased ability of oxygen optake

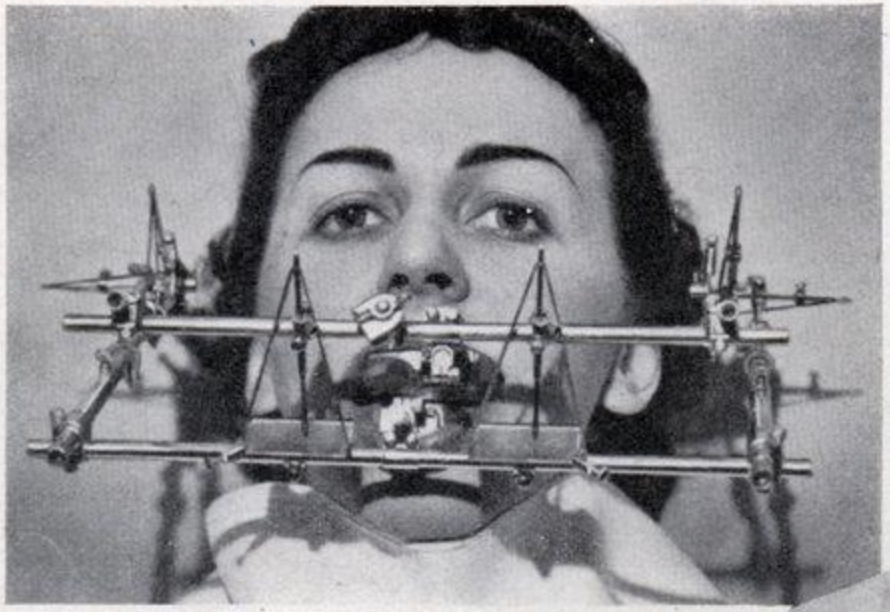




Findings

- 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

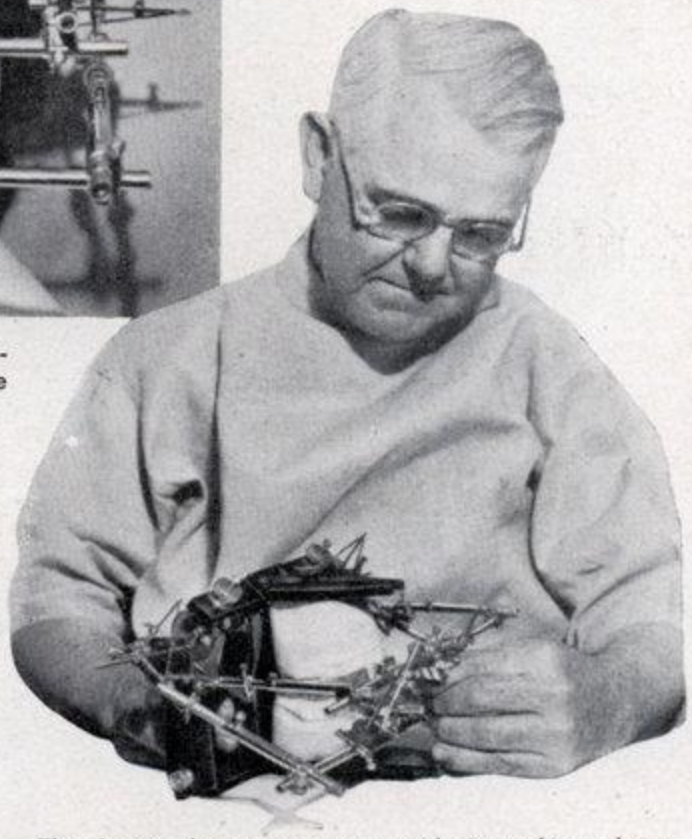
What do we have to measure in patients with lung fibosis??



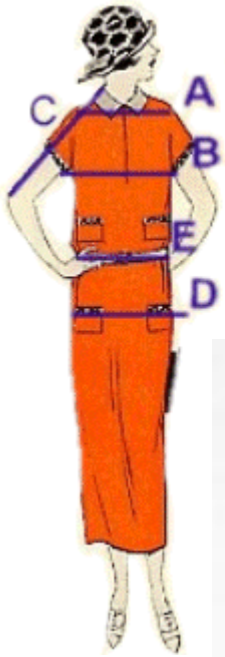
Fitted to a patient's jaws, the "gnathograph" registers the arrangement of teeth and direction of bite

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

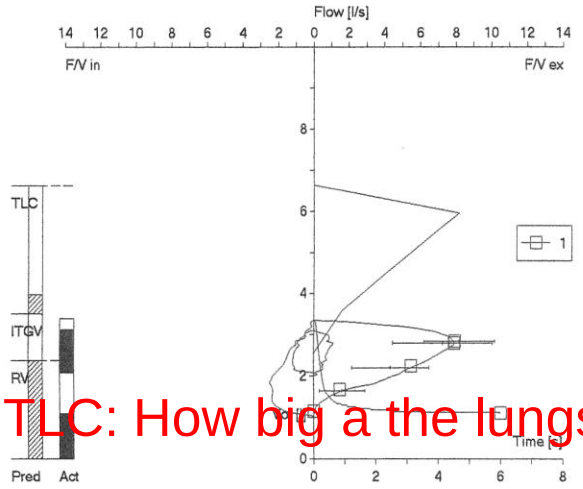
Always initially do TLC;RV and DLCO

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

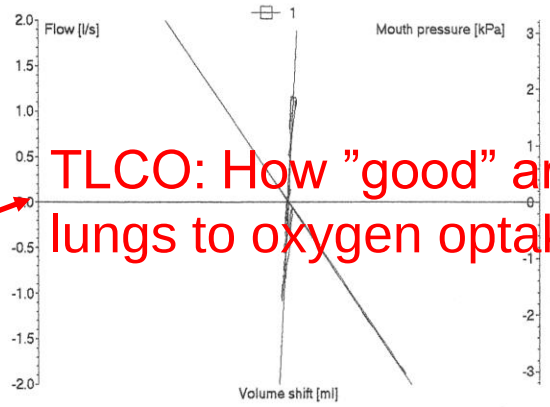
UDVIDET LUNGEFUNKTIONSUNDERSØGELSE

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 ?

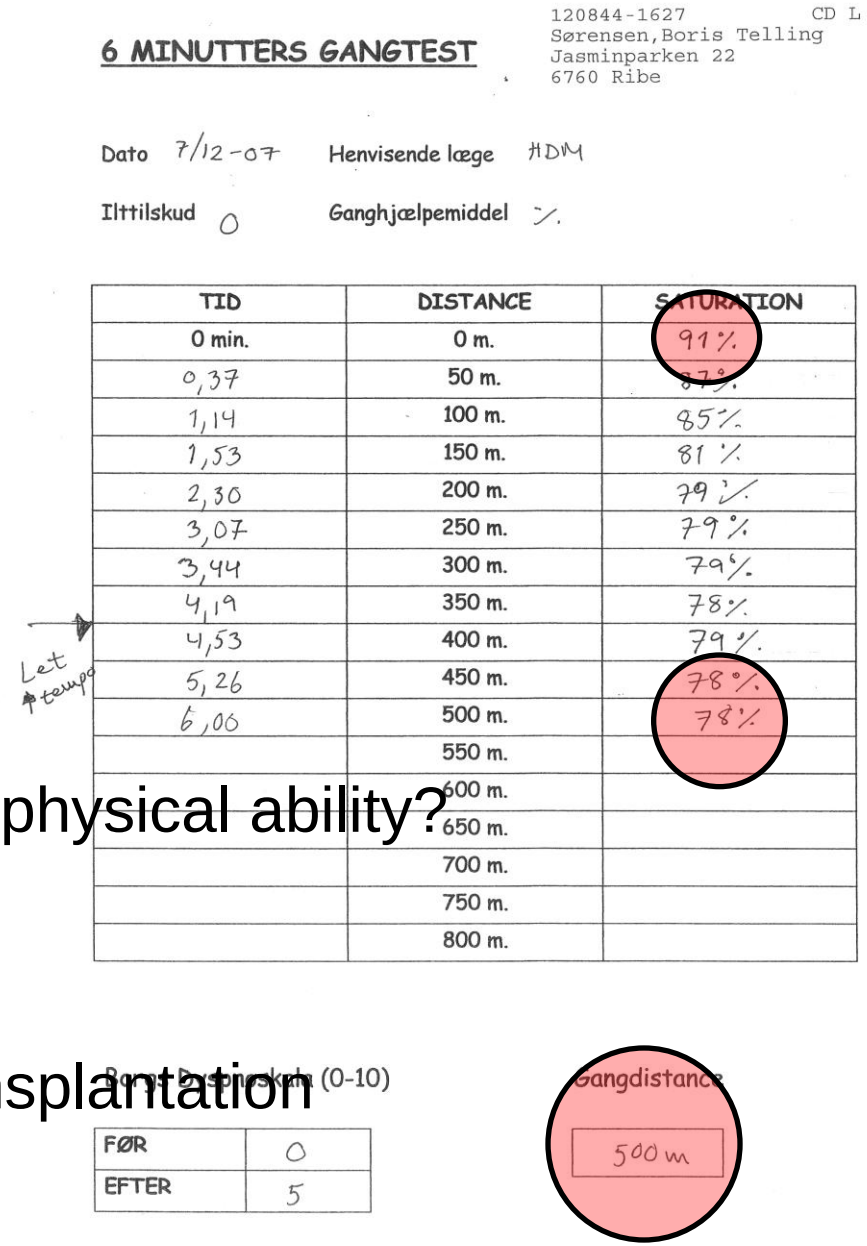


TLCO: How ”good” are the lungs to oxygen optake ?

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

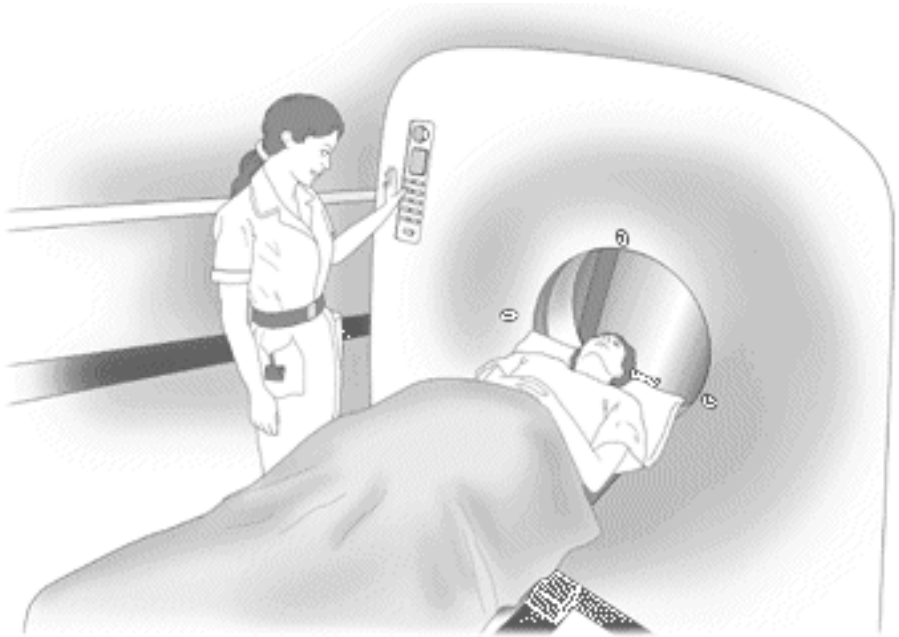
Mette Aukjær

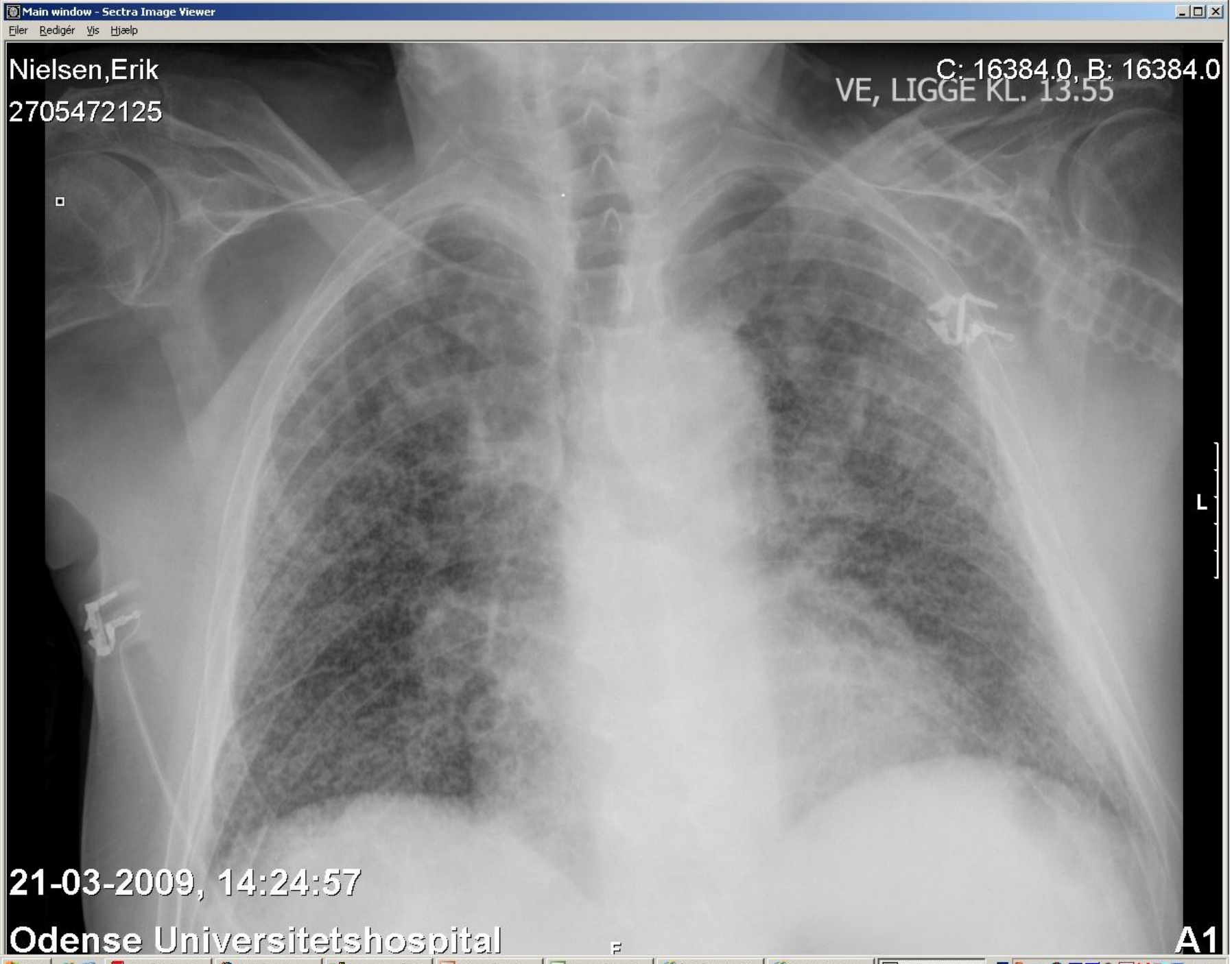
HRCT- scan

- Changes
- distribution
- Pattern

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

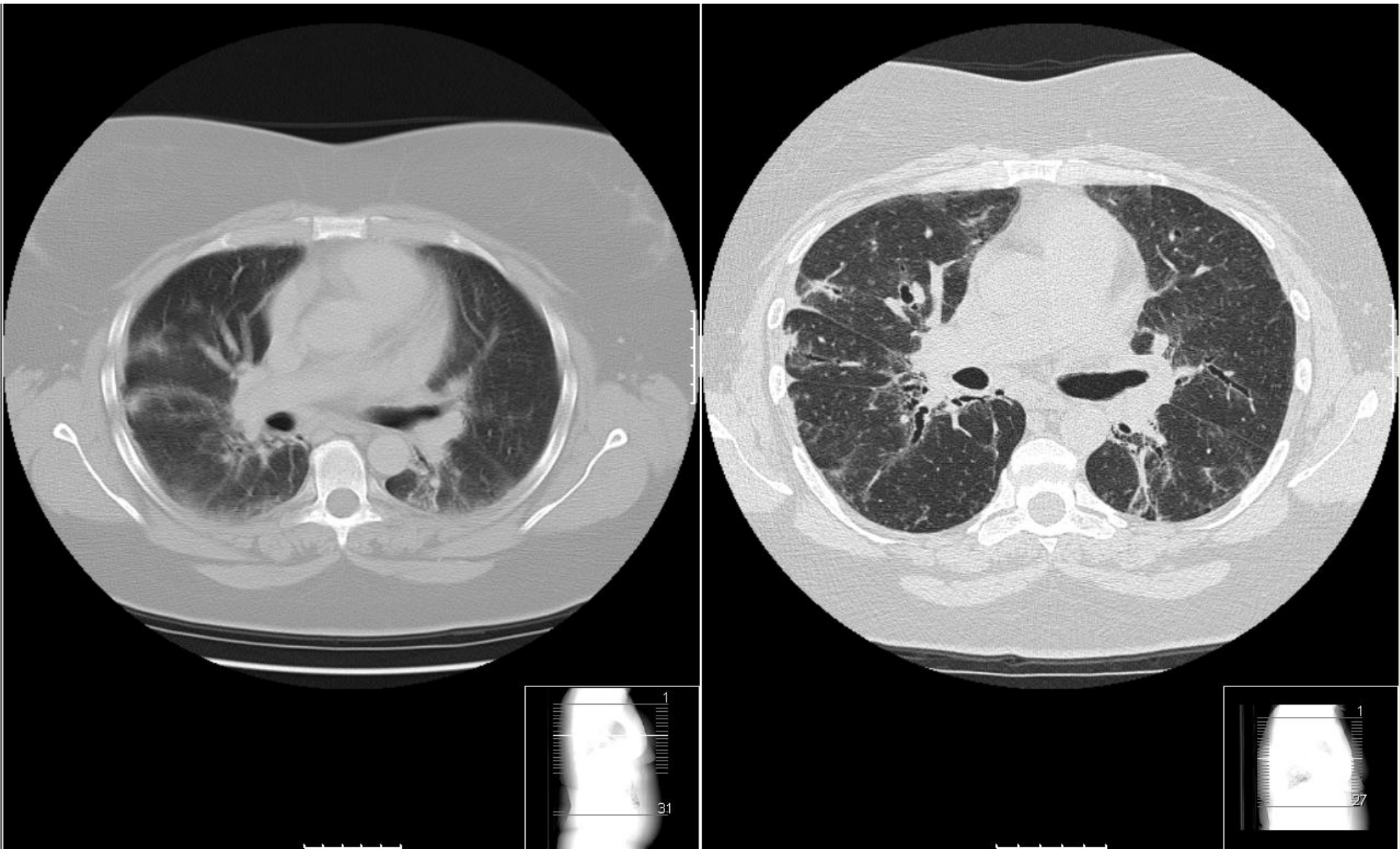
- development
  - Effect of treatment
  - Changes in disease



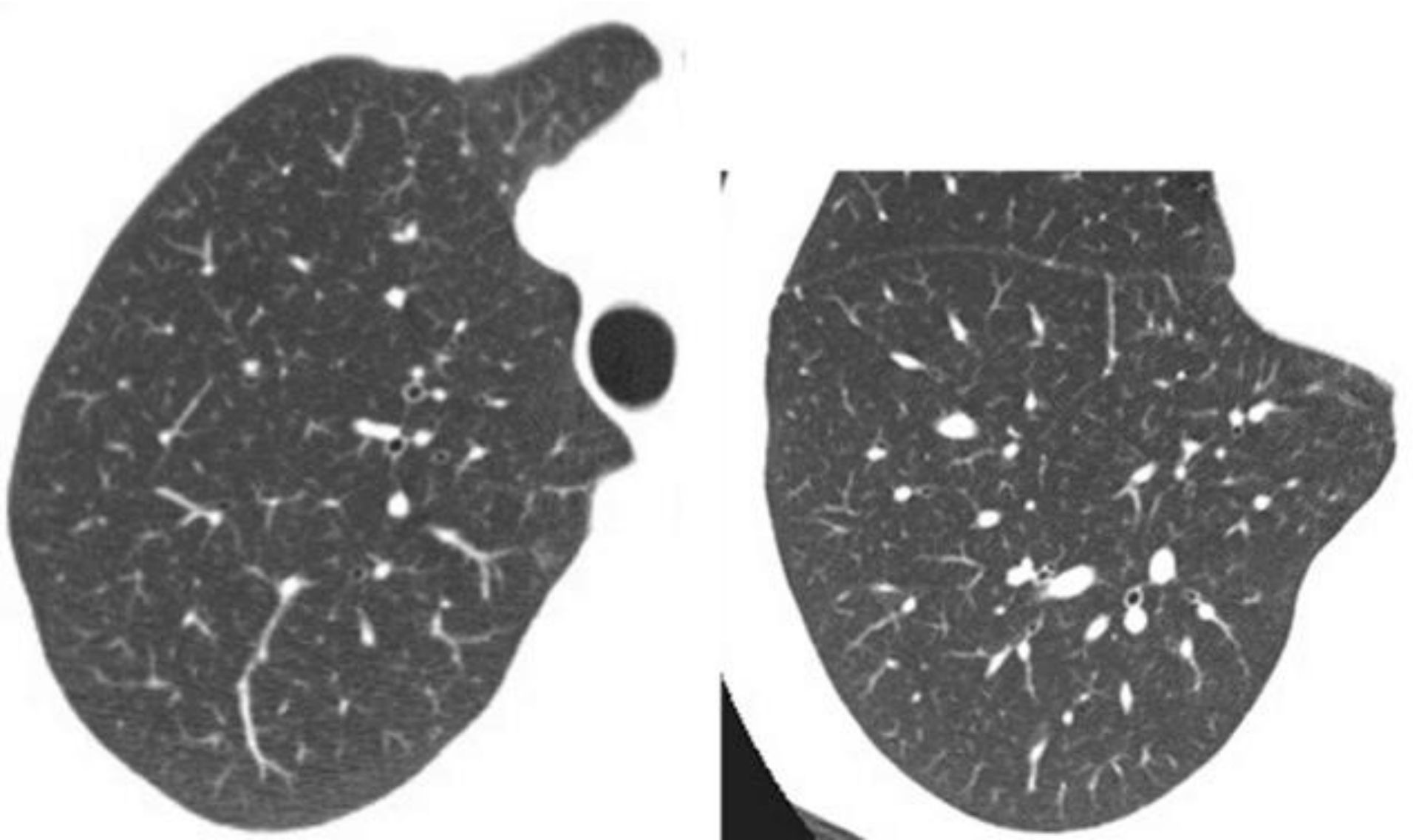

Main window - Sectra Image Viewer
Filer Redigér Vis Hjælp
Nielsen,Erik
2705472125
C: 16384.0, B: 16384.0
VE, LIGGE KL. 13.55
L
21-03-2009, 14:24:57
Odense Universitetshospital
A1
Start
Novell-delivere...
Novell GroupW...
Session A - [24...
Microsoft Pow...
Microsoft Excel...
fpn.dk - Miss U...
PACS Gateway...
Main window ...
12:38

CT versus HRCT

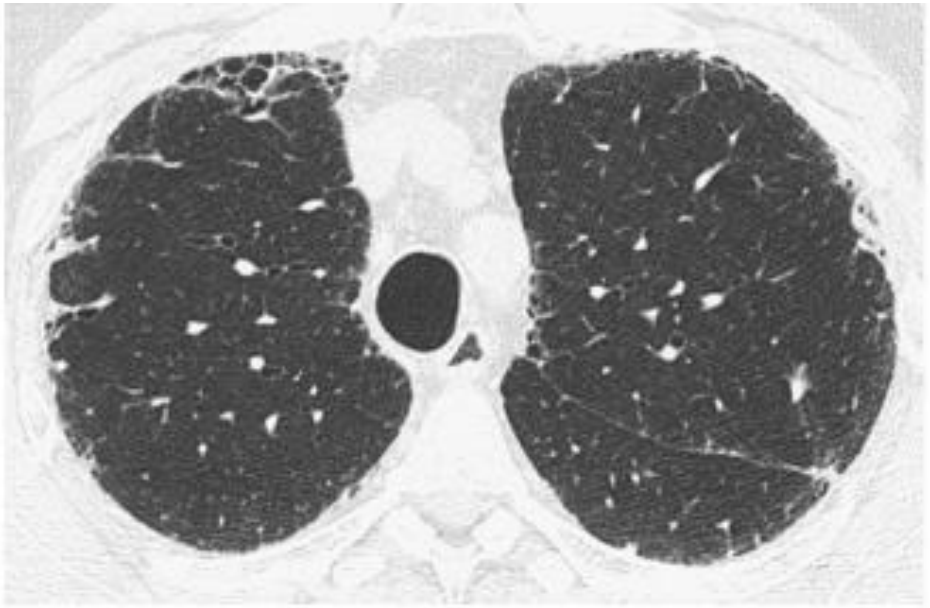
Always HRCT !!!

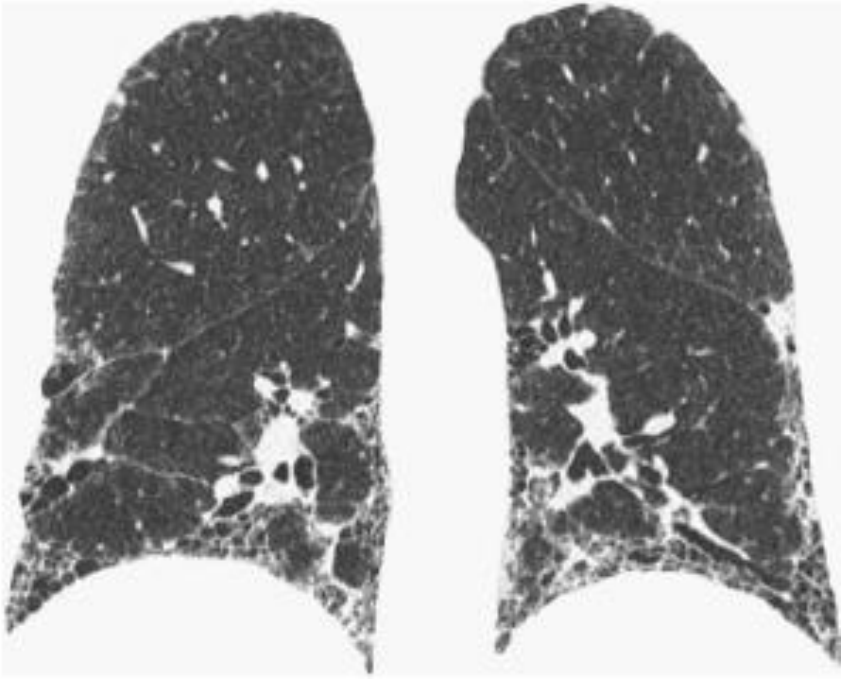


HRCT normal lung



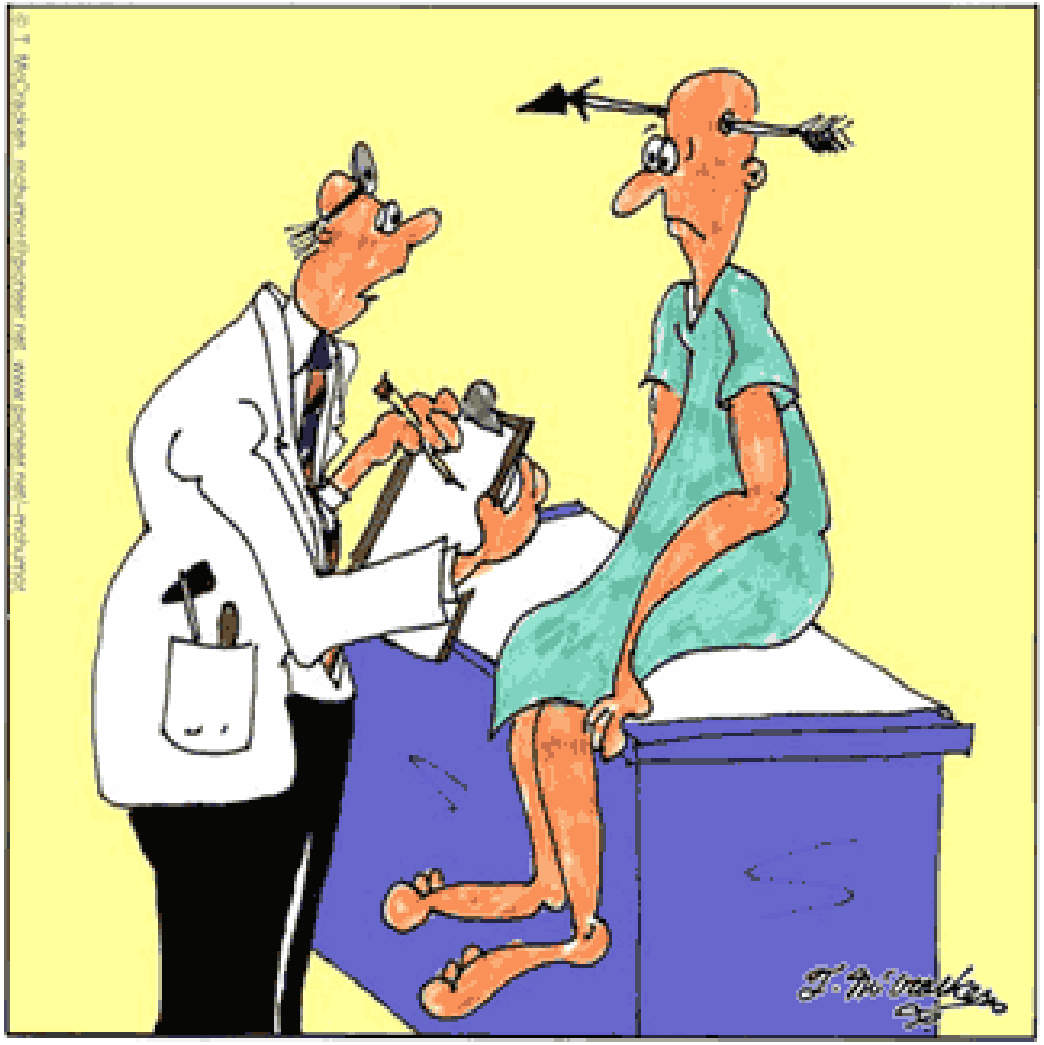
HRCT in a patient with lungfibrosis (UIP)



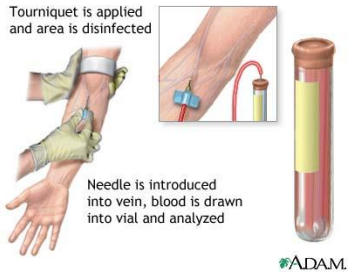


Bloodtests

- Diagnosis
- Prognosis



"Off hand, I'd say you're suffering from an arrow through your head, but just to play it safe, I'm ordering a bunch of tests."



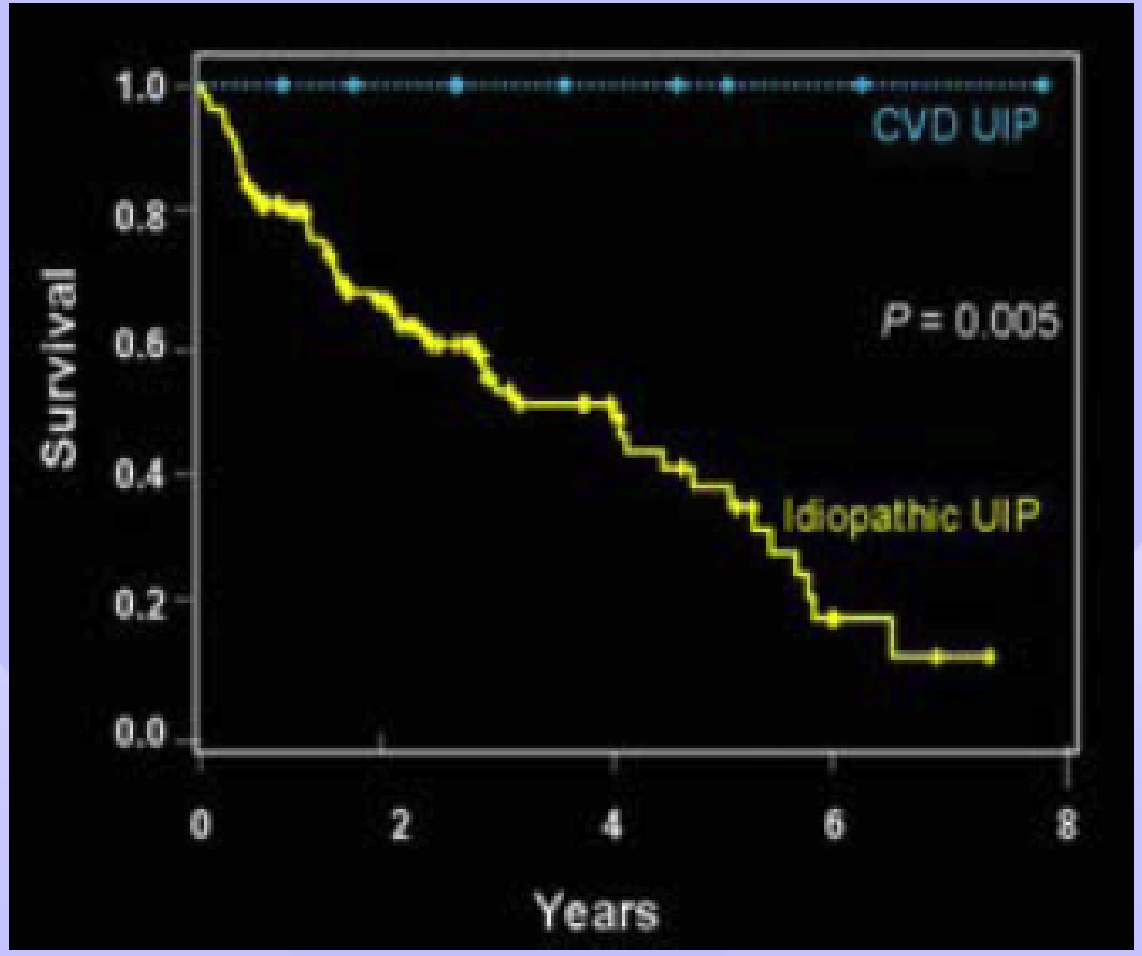
Bloodtests

- Common bloodtests…..
- Extra Interstitial blood tests:
  - ACE: Sarcoidose
  - ANA: Relaterede to ex - (SLE,RA)
  - ANCA: Wegeners granulomatosis
  - Anti-CCP Reumatoid artritis
  - IgM-RF Reumatoid artritis
  - GBA Goodpasteurs disease
  - SLC-70 Sclerodermi
- Arterial-gas

Collagen-Vascular Disorders

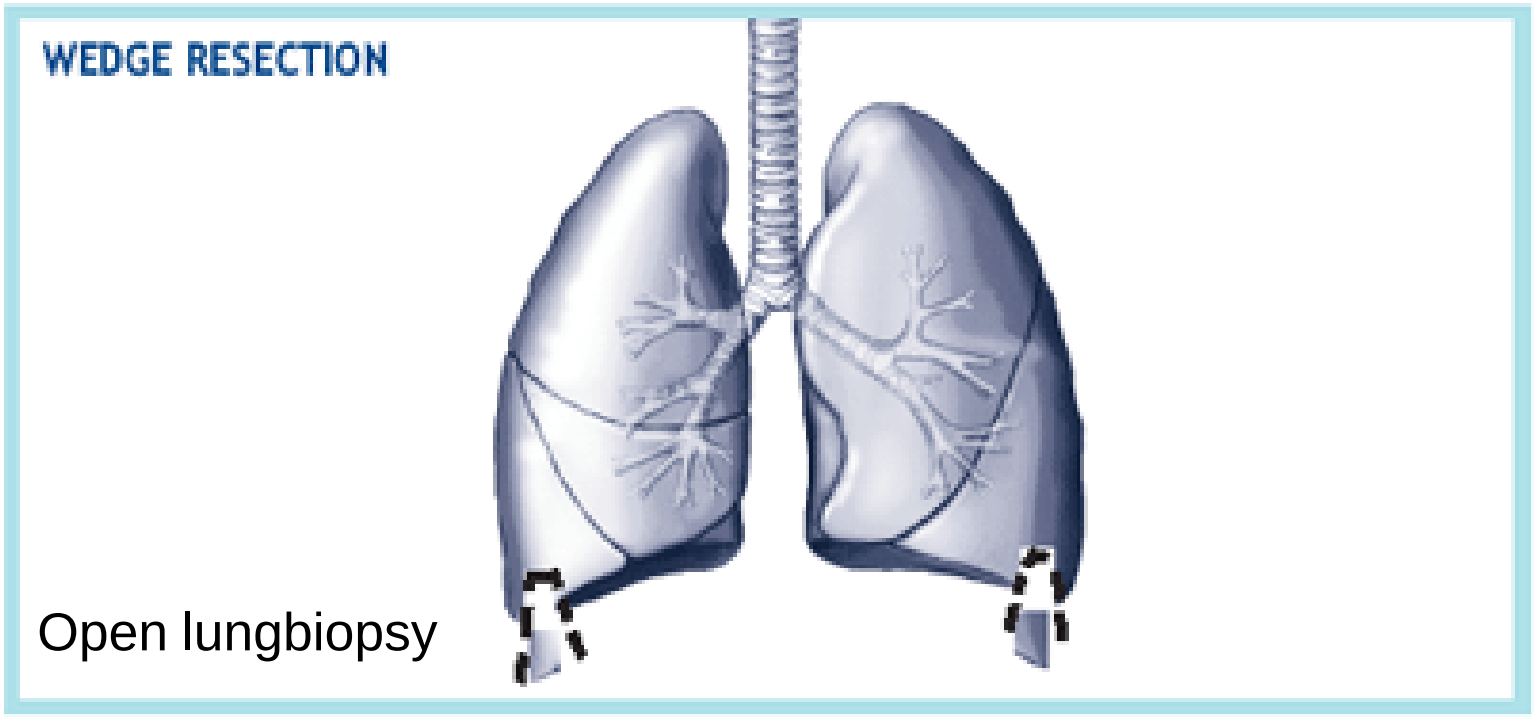
UIP due to underlying collagen vascular disease is associated with a much better prognosis than idiopathic UIP (IPF)

- usual treatment consists of standard immunosuppressive regimens.



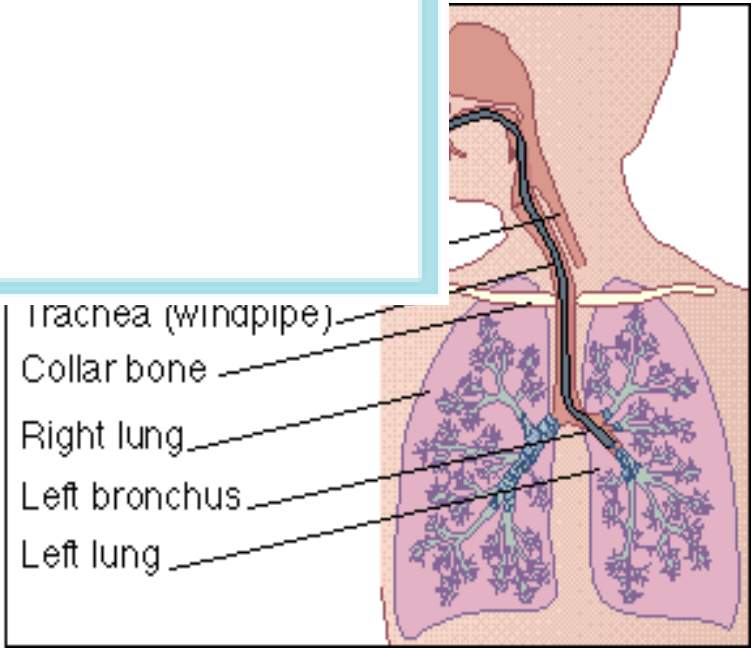
Flaherty KR, et al. *Am J Respir Crit Care Med*. 2003;167:1410-1415.

Bronchoscopy



Open lungbiopsy

is



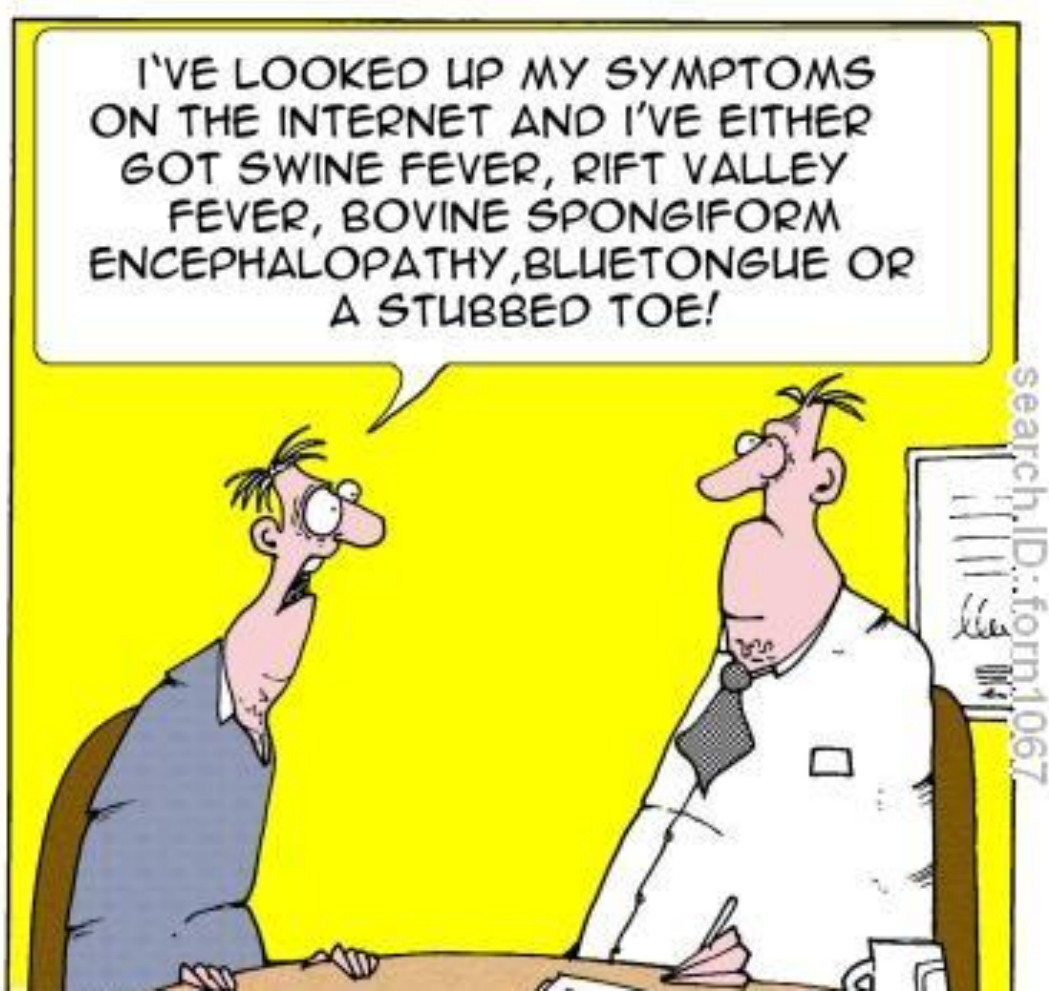
And then!

Think!

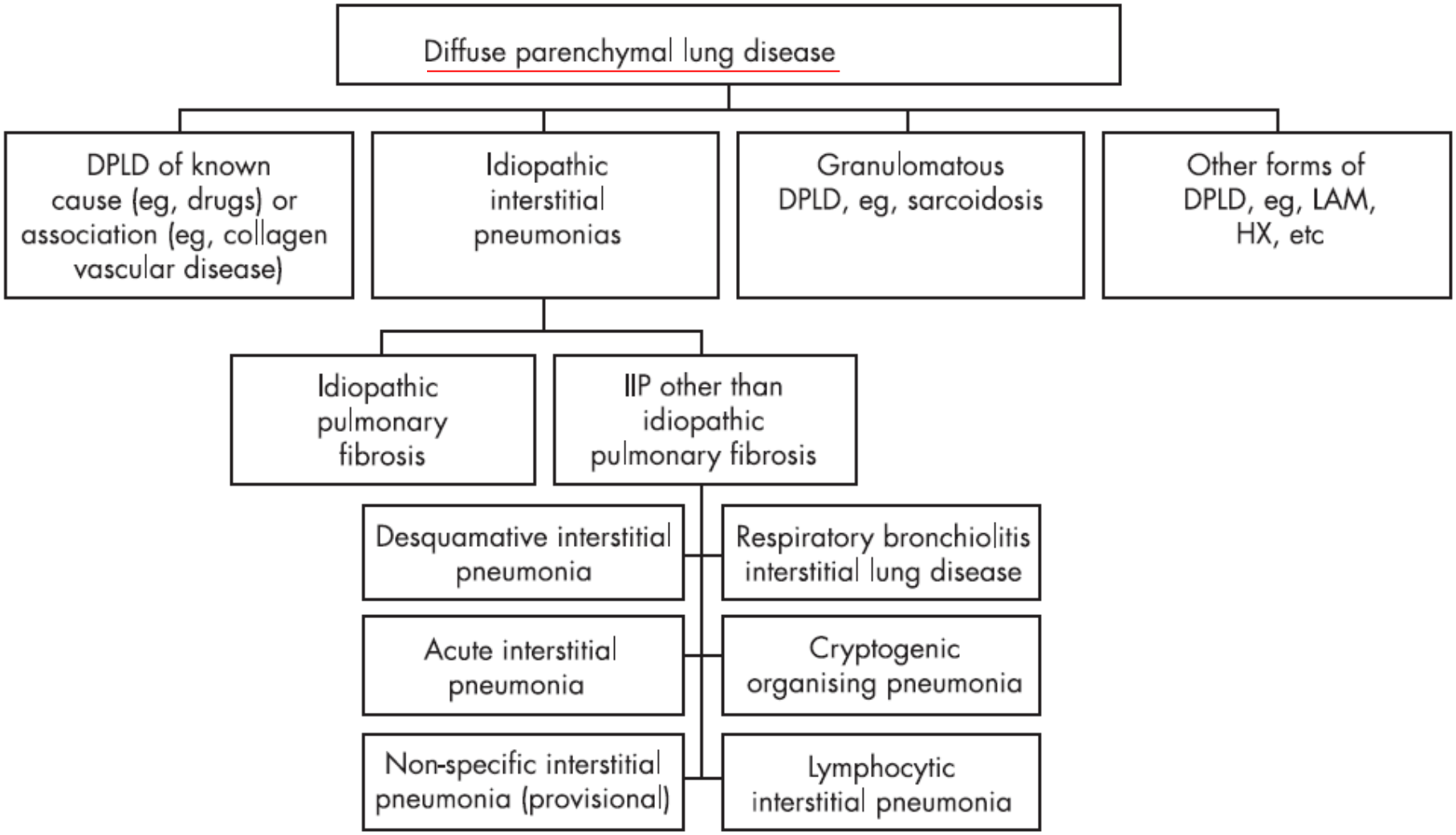
Dignosis !

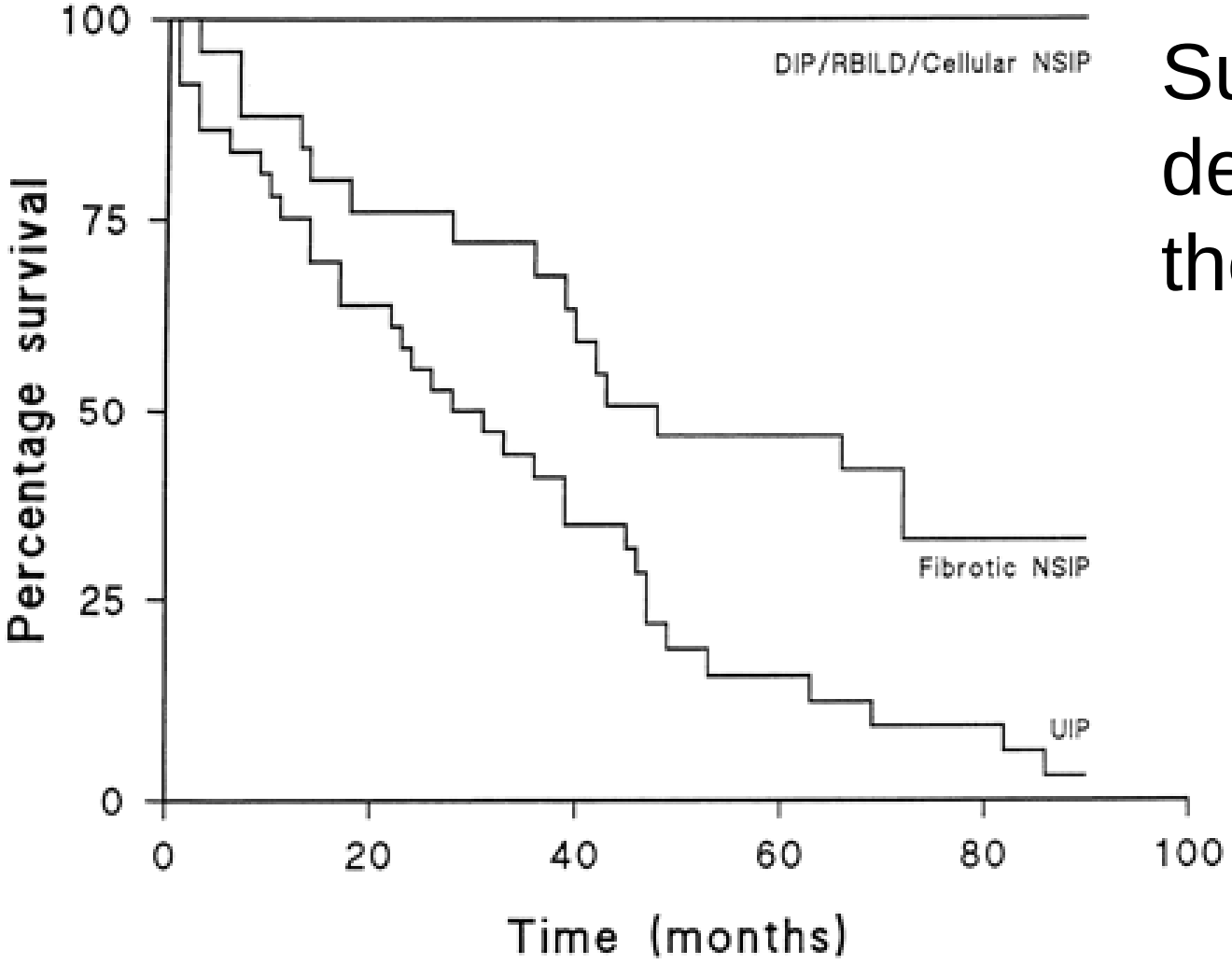
Treatment!’

Remenber ONLY
For lungspecialist!!!



classification





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


LAST
CHANCE

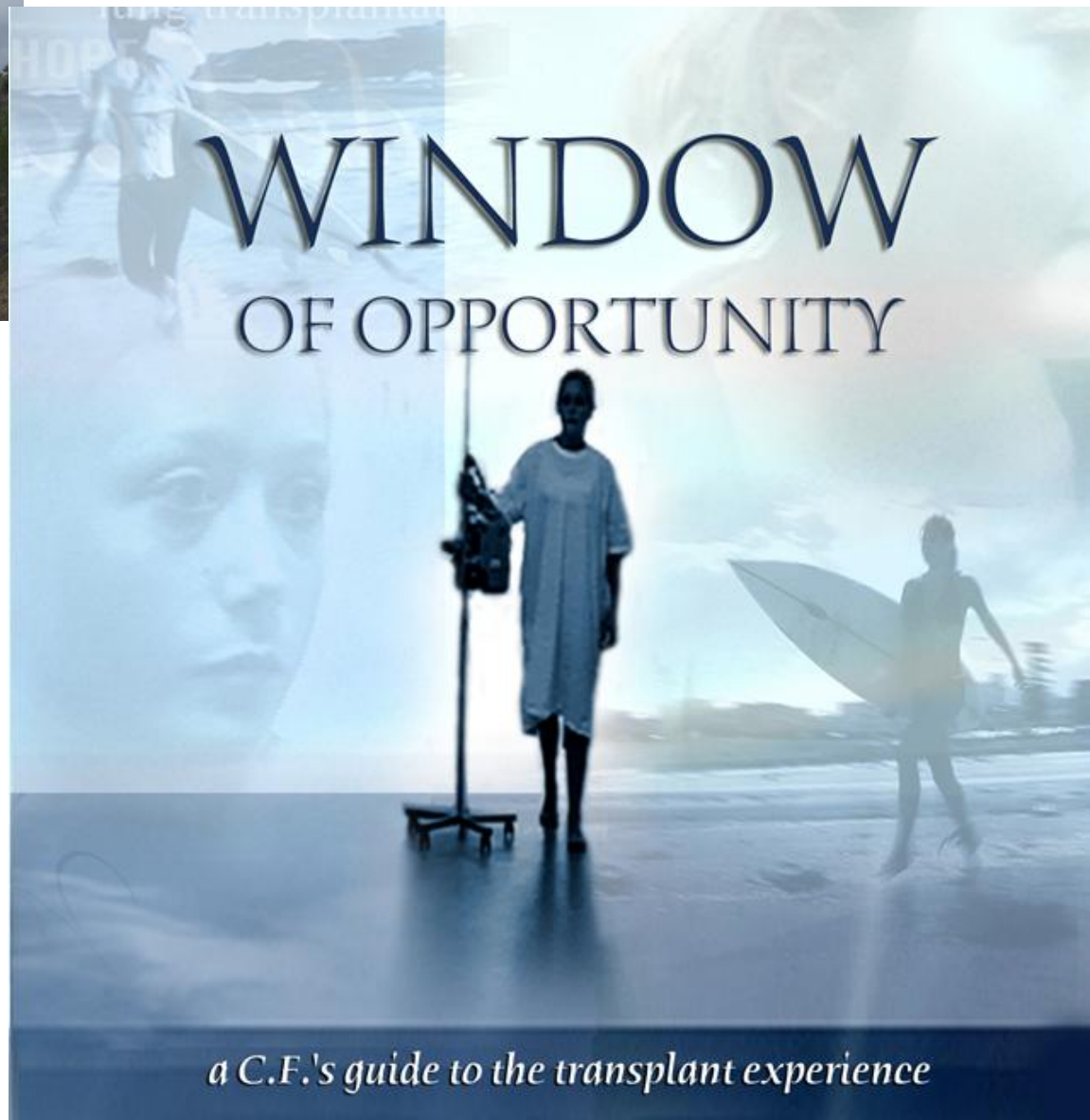

WINDOW
OF OPPORTUNITY
a C.F.'s guide to the transplant experience

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

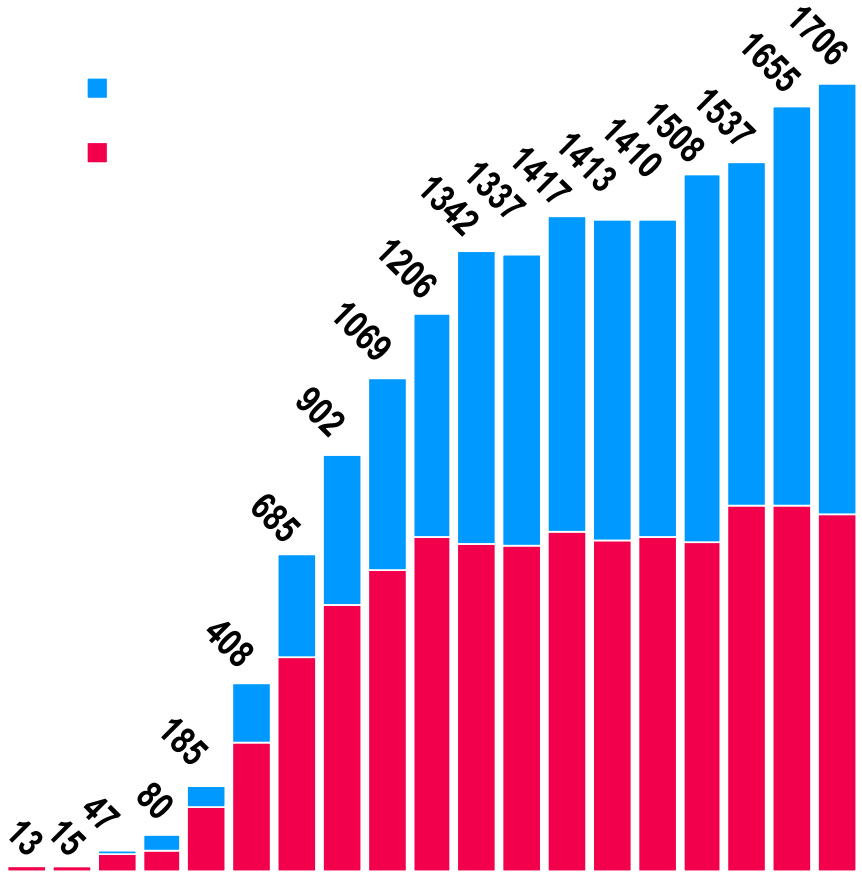
Eisenmenger's syndrome

NYHA functional class III or IV despite optimal medical management

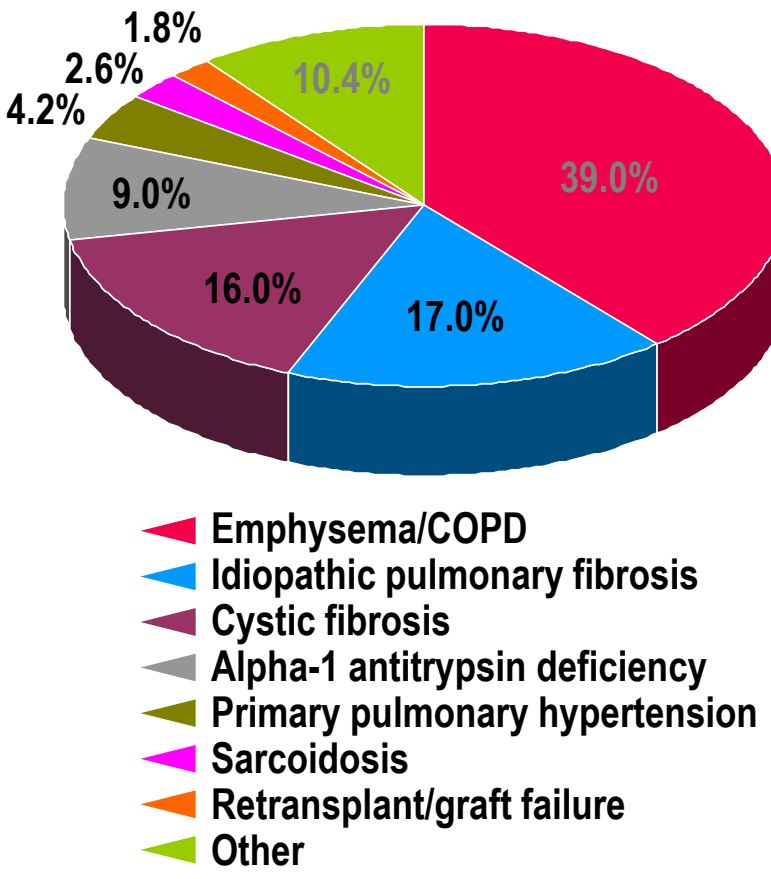
- transplantation is dependent on the disease

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

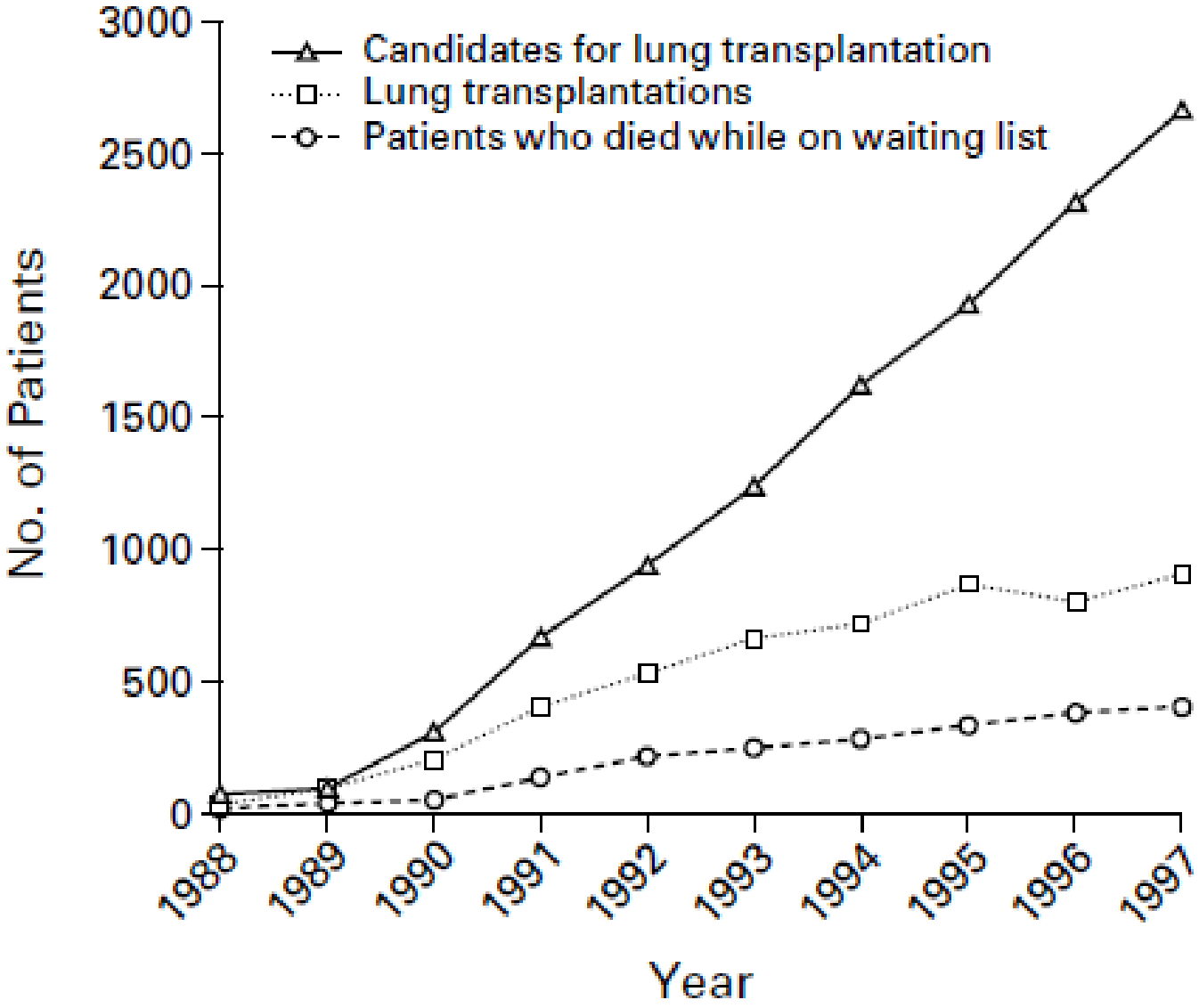


Figure 1. Lung Transplantation in the United States, 1988 to 1997.

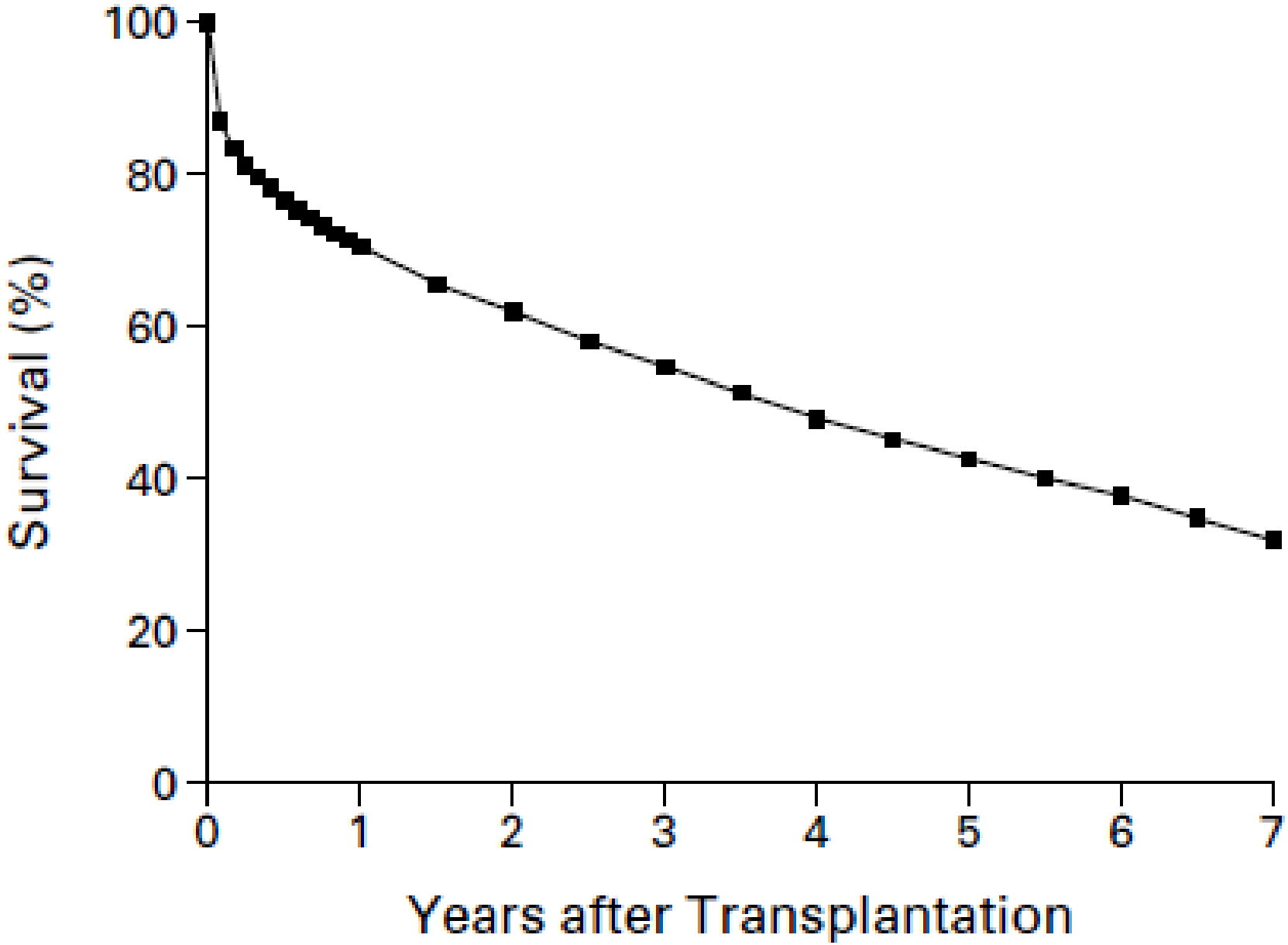
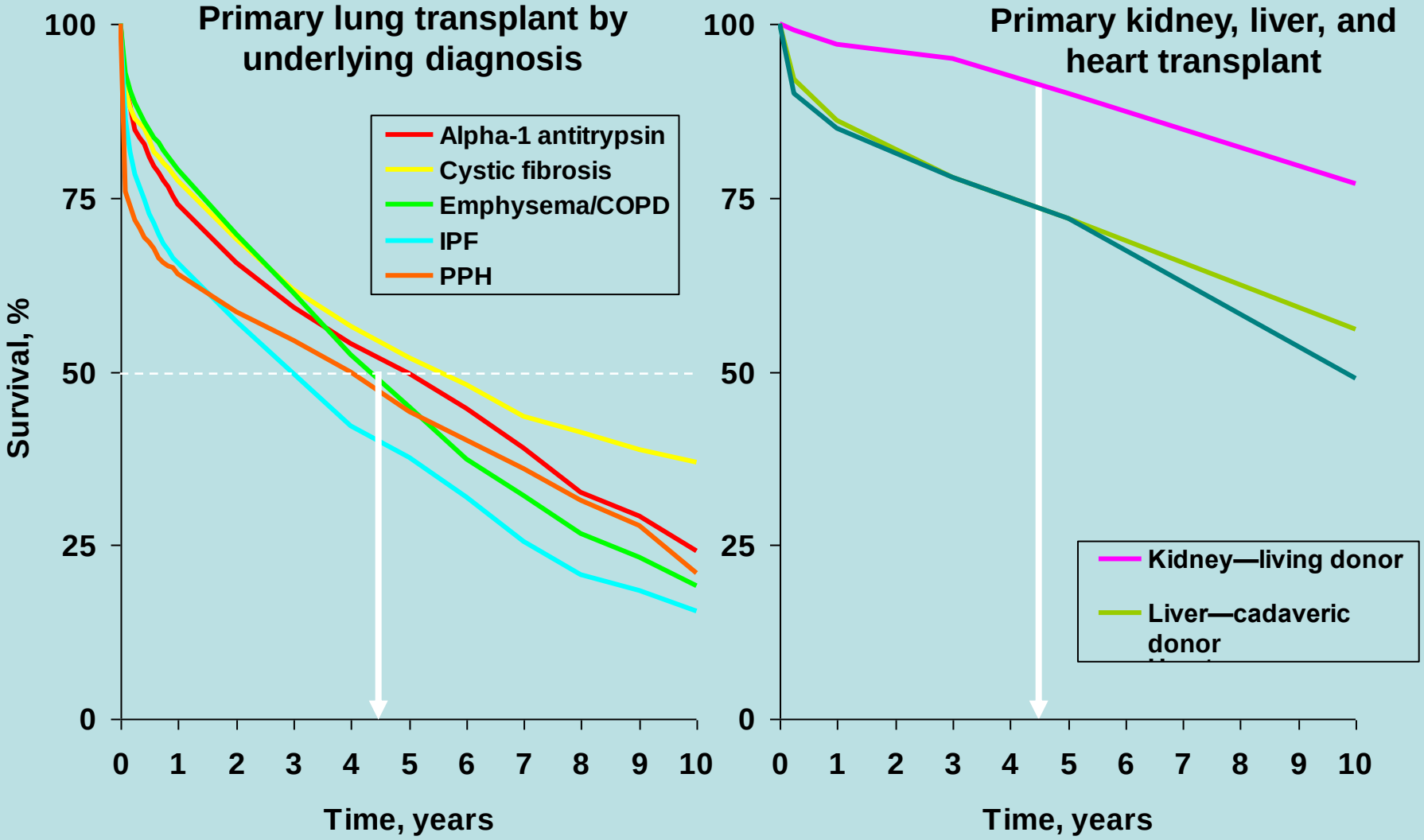


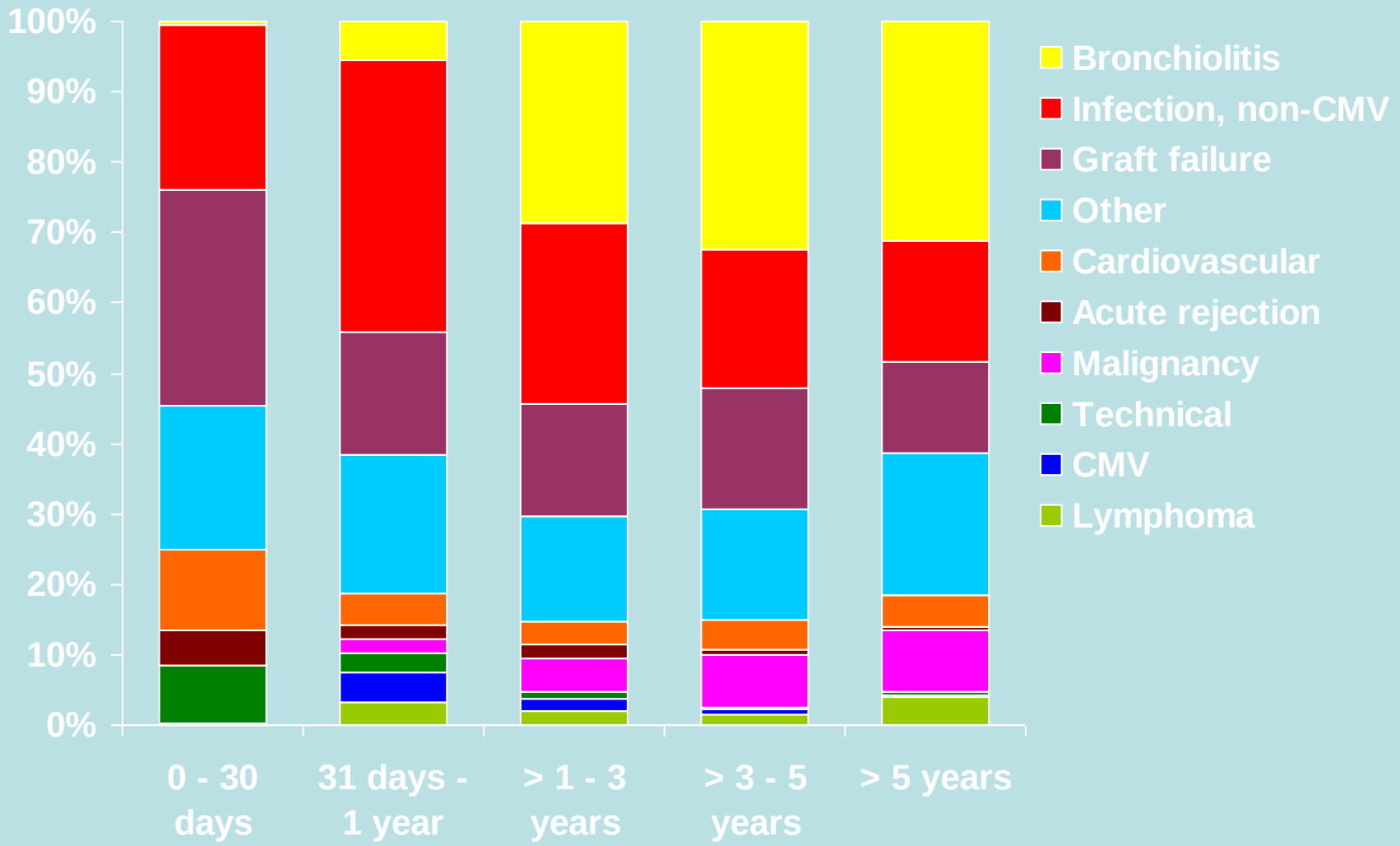
Figure 2. Actuarial Survival after Lung Transplantation.

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???