



**Karolinska  
Institutet**

# **Lungmanifestationer vid myosit**

**20 maj 2025**

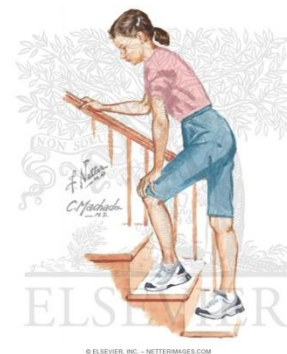
## Föreläsare

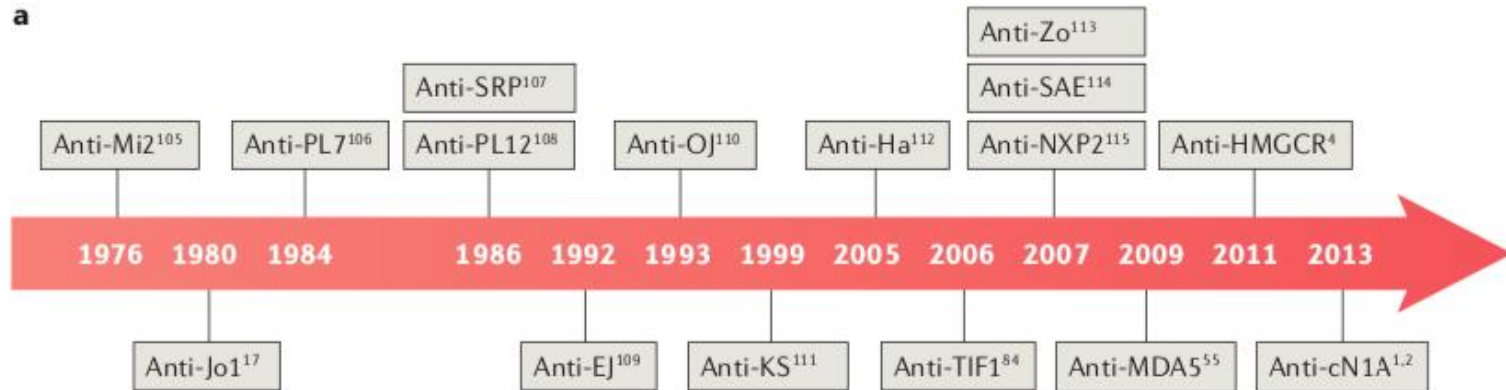
Antonella Notarnicola, MD, PhD, Biträdande överläkare i Reumatologi

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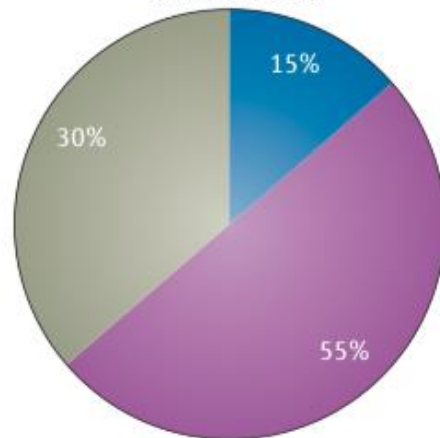
# Myosit = muskelinflammation

- Muskelsvaghet i proximal och nackmuskulatur
- Nedsatt muskelstyrka och muskuläruthållighet
- Inflammation i muskelvävnader
- Stegring muskel enzymer: CK, LD, ASAT
- EMG: myopatisk bild
- MR: muskelödem
- Hud: Gottrons papler, heliotrop exantem
- Sväljningssvårigheter

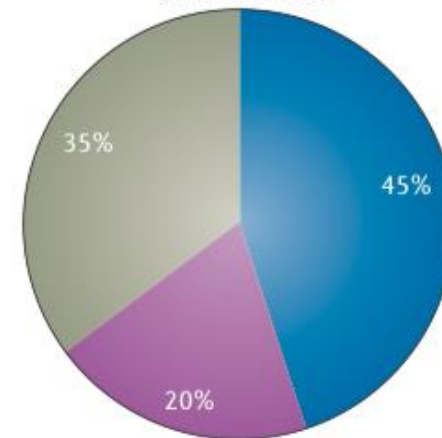




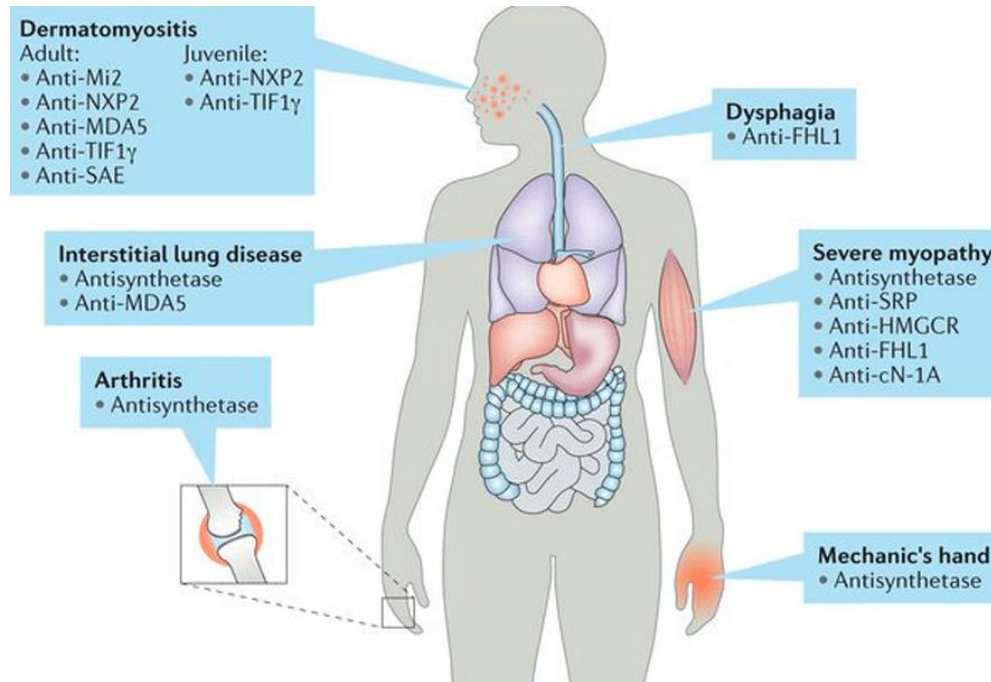
**b** MSAs or MAAs in juvenile-onset myositis (estimated %)



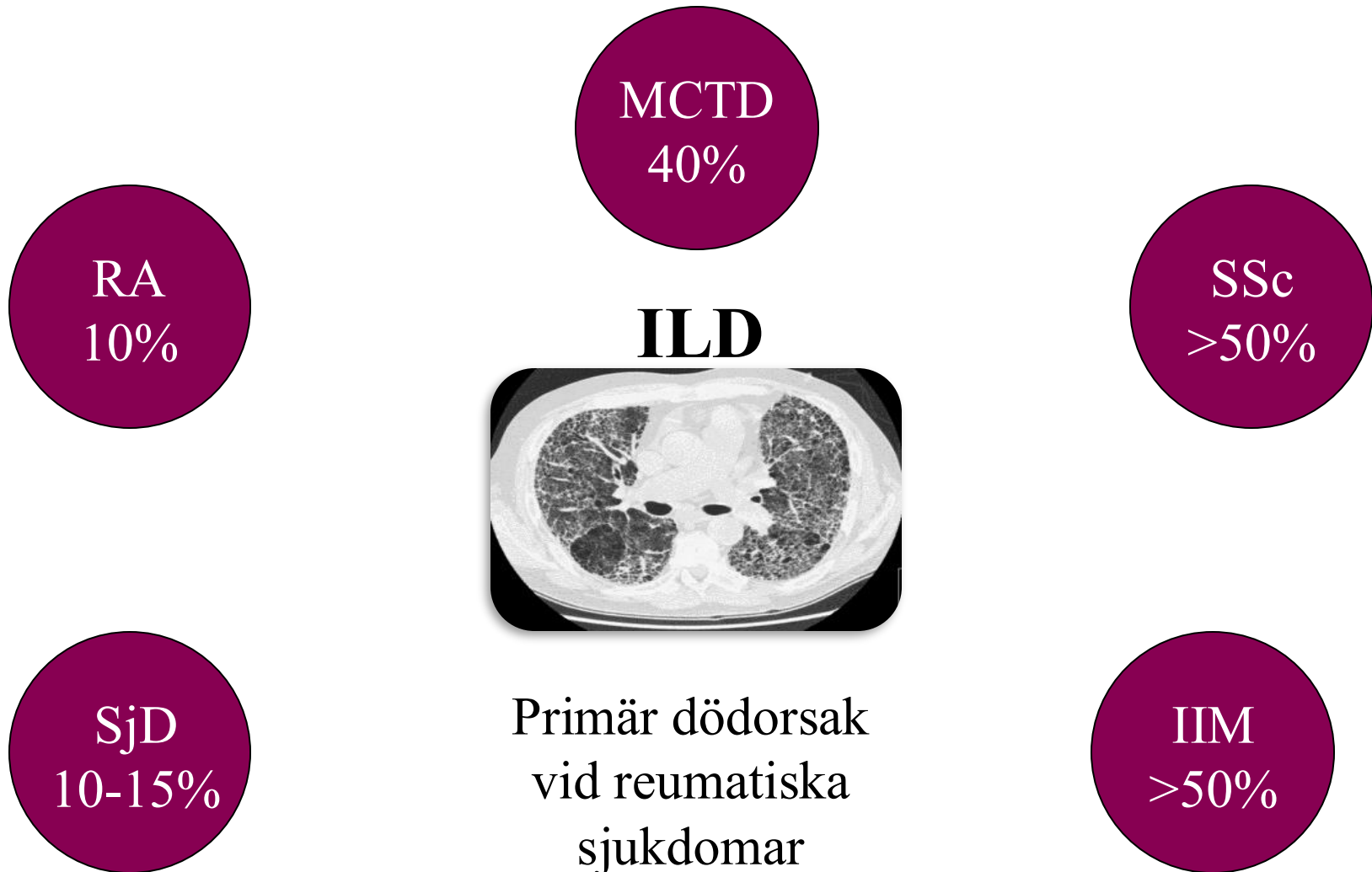
MSAs or MAAs known in adult-onset myositis (estimated %)



● Antibodies known pre-2005   
 ● Antibodies discovered since 2005   
 ● Antibodies unknown



- Polymyositis (PM)
- **Dermatomyositis (DM)**
- Clinically amyopathic dermatomyositis (CADM)
- Immune-mediated necrotizing myopathy (IMNM)
- Cancer-associated myositis (CAM)
- Overlap myositis (OM)
- **Antisynthetase syndrome (ASSD or Asys)**
- Inclusion body myositis (IBM)



## Om lungfibros

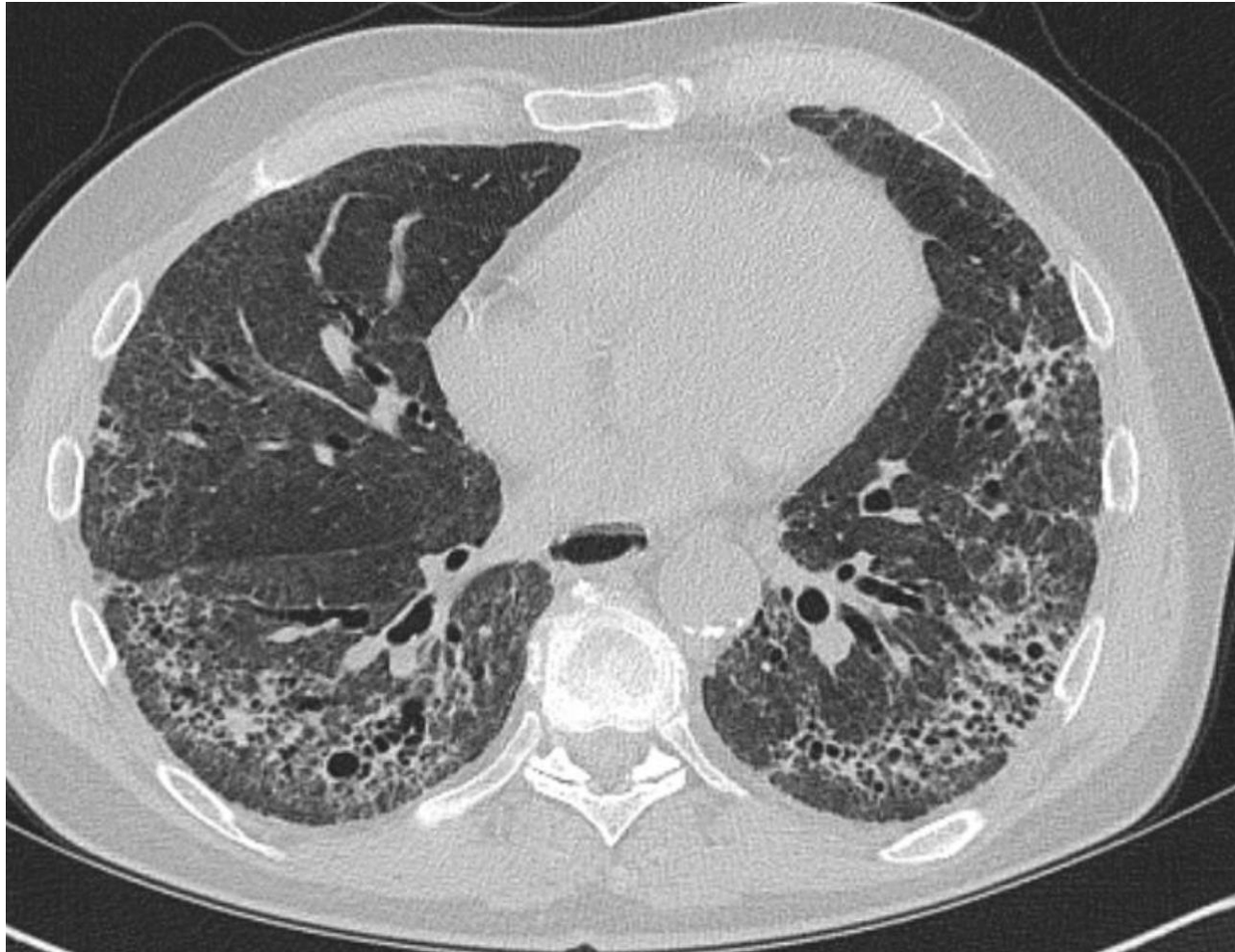
**Interstitiell lungsjukdom** (interstitial lung disease, ILD) är ett begrepp som beskriver en heterogen grupp av lungsjukdomar. ILD drabbar båda lungorna i varierande grad och omfattar ett stort antal tillstånd med ett brett spektrum av orsaker.

**Lungfibros** är ett **samlingsbegrepp** för tillstånd med ärrömvandling av lungvävnad och ingår i gruppen interstitiell lungsjukdom. Lungfibros utvecklas inte hos alla patienter med ILD. Fibrotisering är en skada som inte är reversibel.

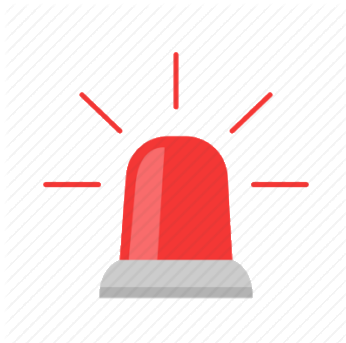
**Idiopatisk lungfibros (IPF)** är den fibrotiserande lungsjukdom som är mest beskriven och karakteriserad.

**Den radiologiska bilden med högupplöst datortomografi (HRCT)** är en hörnsten i diagnosticering av ILD och särskilt vid fibrotiserande lungsjukdom.

# NSIP-non specific interstitial pneumonia



- Peripheral reticulation
- ground-glass
- Traction bronchiectasies
- Subpleural sparing
- Bilateral involvement



Symtom: andfåddhet, torrhosta  
Lungauskultation: krepitationer



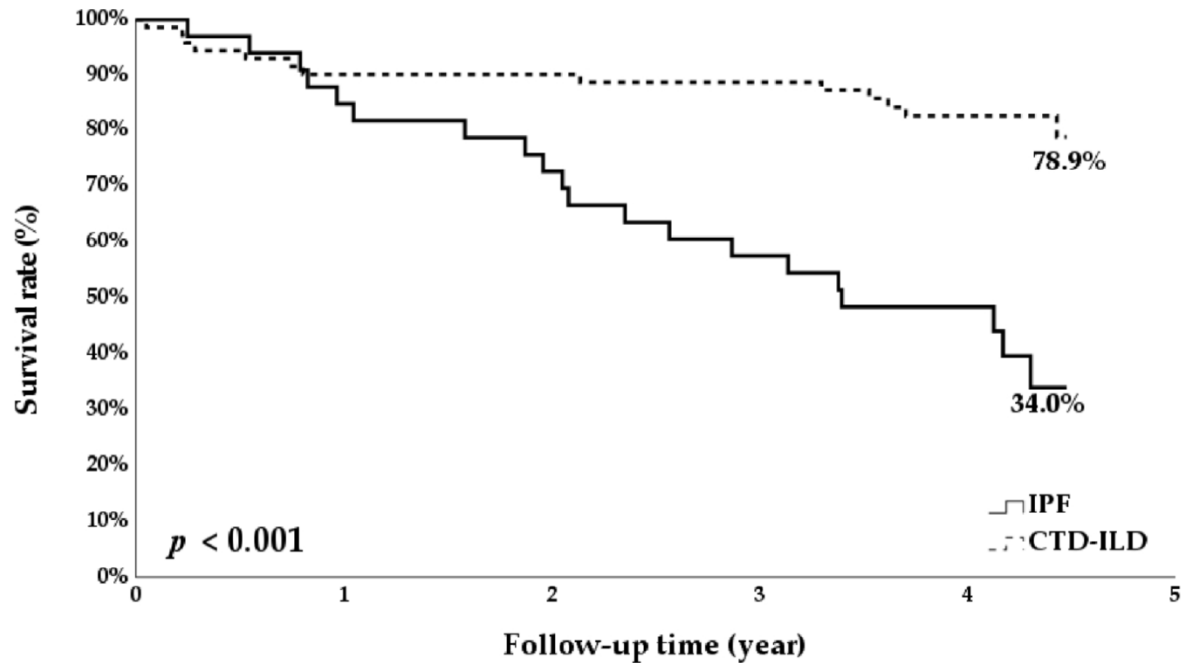
- Nedsatt lungfunktion: FVC eller TLC <80% av det förväntade värdet
- Minskad diffusion av kolmonoxid (DLCO)
- FVC <60% har associerats med dålig prognos

**OBS**

**ILD kan hittas även även hos asymptomatiska patienter**



# Survival of CTD-ILD vs IPF

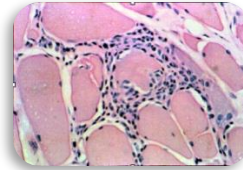
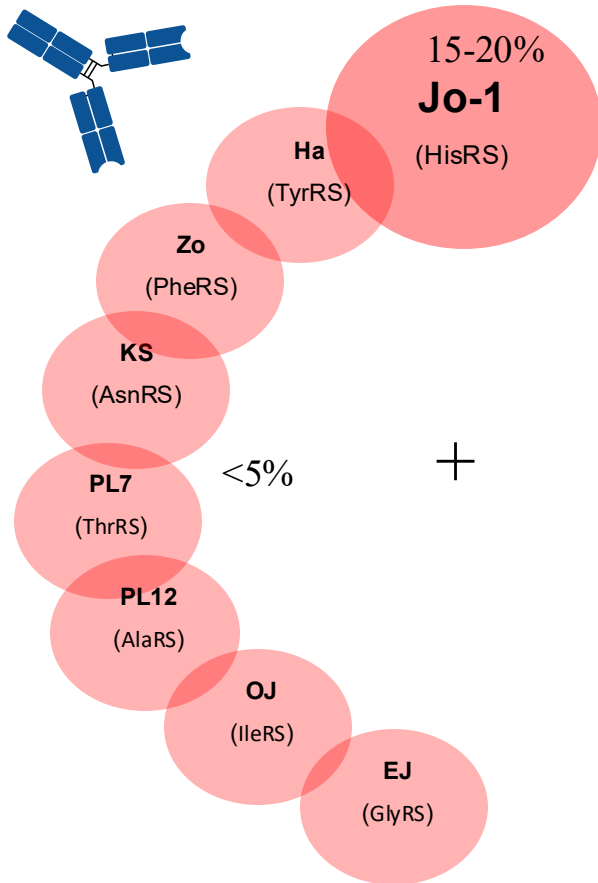


Case Processing Summary

	Total	Dead	Alive	Alive Percent	Survival rate (%)					<i>p</i> for log rank
					1 year	2 year	3 year	4 year	5 year	
IPF	33	20	13	39.4%	84.8%	72.7%	57.6%	48.5%		<0.001
CTD-ILD	71	13	58	81.7%	90.1%	90.1%	88.7%	82.7%		
Overall	104	33	71	68.3%						

Kaplan–Meier survival curve according to the interstitial lung disease condition

# Antisyntetas syndrom (ASSD or ASyS)



Myosit  
(PM/DM)



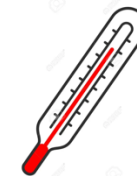
Artrit



Interstitiell  
lungsjukdom  
(ILD)



Mechanics'  
hands



Feber



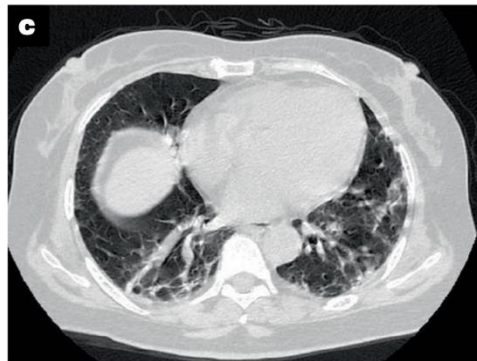
Raynaud  
fenomen

# Anti-MDA5+ dermatomyositis



Prevalens av ILD: 44-100%

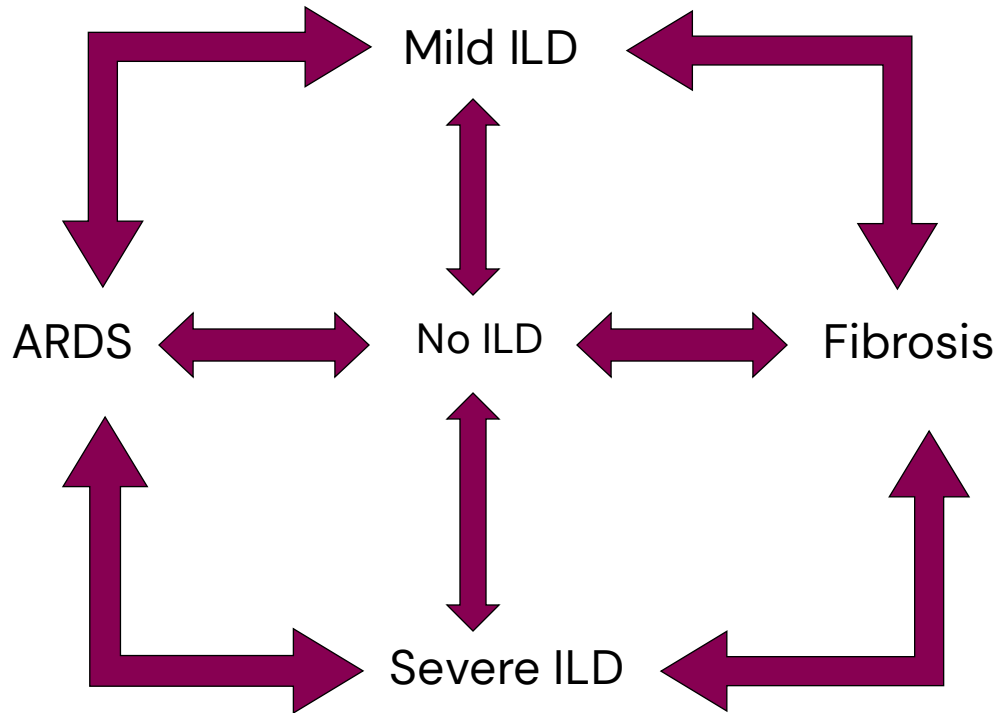
Hög risk för RP-ILD (snabbt progredierande ILD) och hög mortalitet fram för allt hos asiatiska patienter



# Nytt projekt



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*What predicts each patient's direction?*

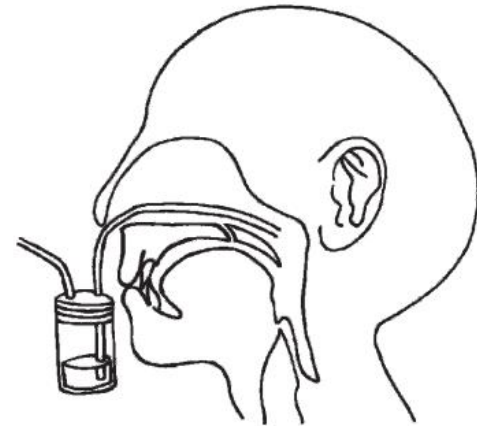


Courtesy of Irene Peralta and Deniz Demirdal

## Pilot projekt:

Non-invasive approach for detection of autoantibodies and T cells in the respiratory compartment of patients with myositis-associated **ILD**

- **Research aim:**
  - Identify anti-Jo1/MDA5 and specific T cells in nasopharyngeal aspirate (NPA) and induced sputum from patients with IIM-related ILD.



**Figure 1** Nasopharyngeal aspiration.

Wai AK et al. Patients' perceptions of nasopharyngeal aspiration in the emergency department of a teaching hospital in Hong Kong. *Emerg Med J.* 2007 Jan;24(1):35-6.

## Doktorandprojekt:

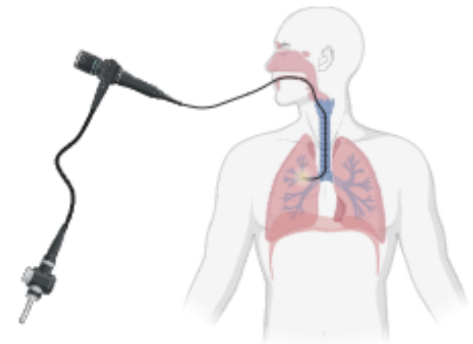
# Phenotyping T cells in blood and lung derived samples of patients with ASSD

- **Research question:**

- To identify and characterize antigen-specific T cells in patients with ILD and ASSD from different compartments: At the time of diagnosis and longitudinally.

- Blood
- BALF
- NPA
- Induced sputum

- What is the T cell phenotype in relation to the ILD pattern, ILD severity and response to treatment?



# TACK!



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