Systemic Sclerosis Interstitial Lung Disease (SSc-ILD):

The Importance of Early Diagnosis, Patient Centered Communication and Evidence Based Treatment

This activity is supported by an independent educational grant from Boehringer Ingelheim Pharmaceuticals, Inc.

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Disclosures

- **Amy Olson, MD** discloses that she on an Advisory Board for Boehringer Ingelheim, on the Board of Directors for MedGraphics as well as a speaker for Boehringer Ingelheim, Genentech, PeerView, Pilot/France Foundation and Vindico. Amy also conducts research for Boehringer Ingelheim.
- **Virginia Steen, MD** discloses that she is on an Advisory Board Boehringer Ingelheim and a Consultant for Boehringer Ingelheim, CSL Behring and Eicos.
- **Zulma Yunt, MD** discloses that she is a Speaker for Boehringer Ingelheim. She also conducts research for Boehringer Ingelheim.
- **Faculty, Planners and Reviewers:** Michael Mohning, MD, Amen Sergew, MD, Andrea Harshman, MHA, CHCP, CMP-HC, Mandy Comeau, Meghan Brenner, and the patient in the video have no relevant financial relationships to report.

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Learning Objectives

- Apply best practices in diagnosis based on clinical symptoms, pathophysiology and disease course of SSc-ILD.
- Utilize evidence-based decision making in the selection of treatments for patients with SSc-ILD.
- Apply strategies for longitudinal management of SSc-ILD using a multidisciplinary approach and patient-centered communication.

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Chapter 1: Diagnosis, pathophysiology, evaluation

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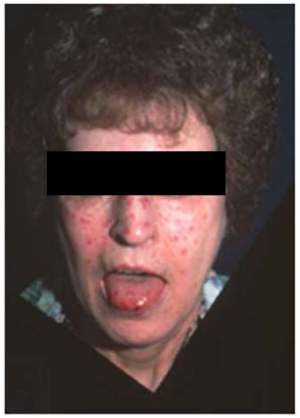
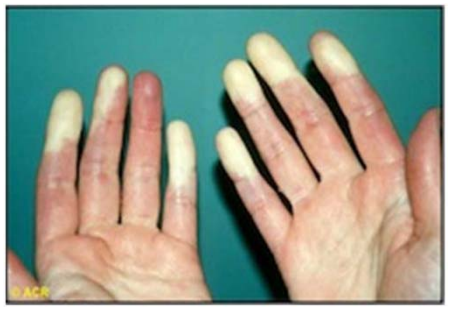
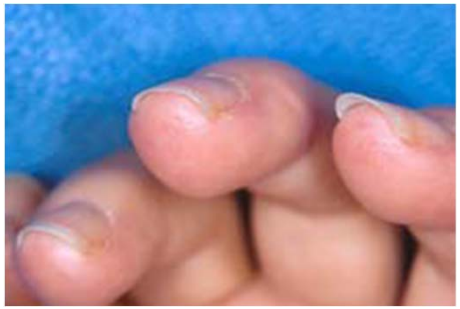
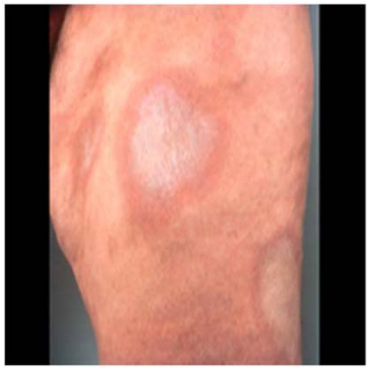
Systemic Sclerosis

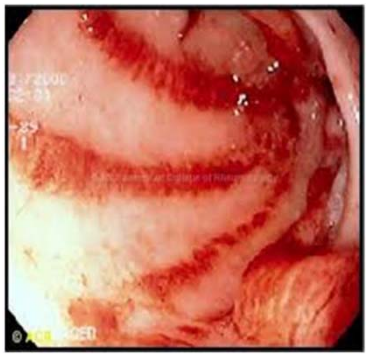
- Systemic, autoimmune, connective tissue disease
- Overproduction and deposition of collagen
- Two predominant forms:
  - Limited cutaneous
  - Diffuse cutaneous
- Lung involvement: ILD and PAH

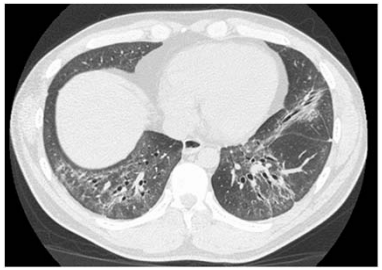












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ACR/EULAR Criteria For Systemic Sclerosis (2013)

3 Hallmarks of SSc
- Vasculopathy
- Auto-antibodies
- Fibrosis of skin and/or internal organs

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers *(only count the highest score)*	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions	Digital Tip Ulcers	2
(only count the highest score)	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerasel [anti-ScL 70], anti-RNA polymerase III)		3
	TOTAL SCORE^:	
Patients having a total score of 9 or more are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

Score ≥ 9 = definite SSc
sensitivity 0.91
specificity 0.92

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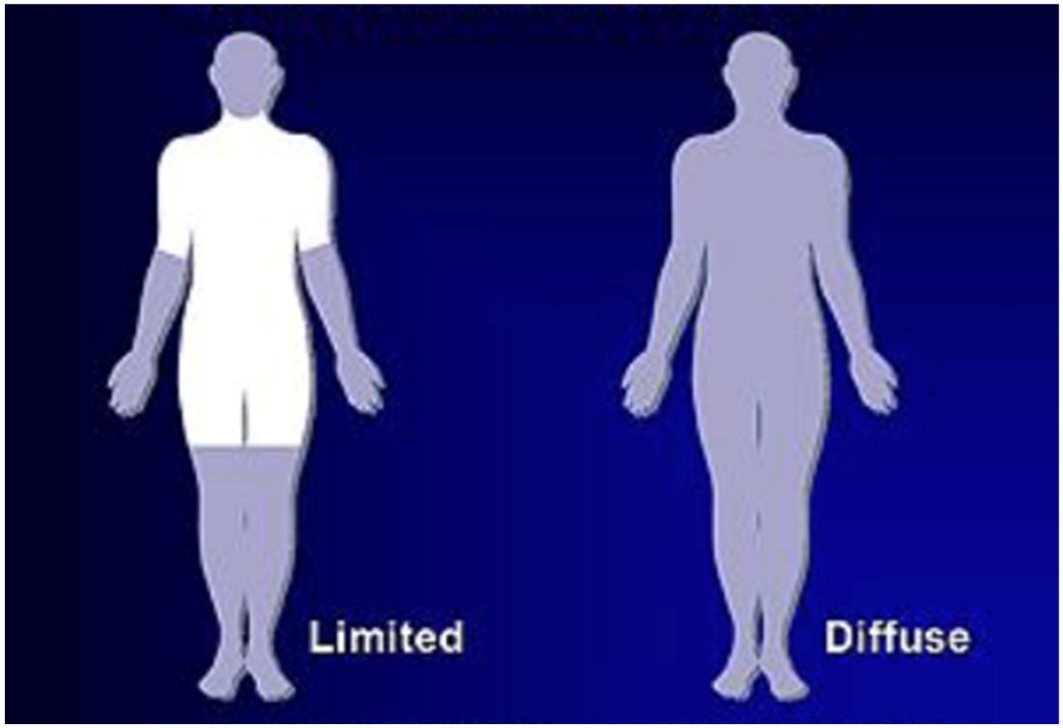
van den Hoogen F, et al. Arthritis Rheum 2013; 65:2737 - Hughes and Pauling. Sem Arth Rheum 2018. 48(5) 888-894

Limited vs Diffuse Cutaneous SSc



Medger T. in Clements and Furst 2nd Ed, Systemic Sclerosis

Limited vs Diffuse Cutaneous SSc

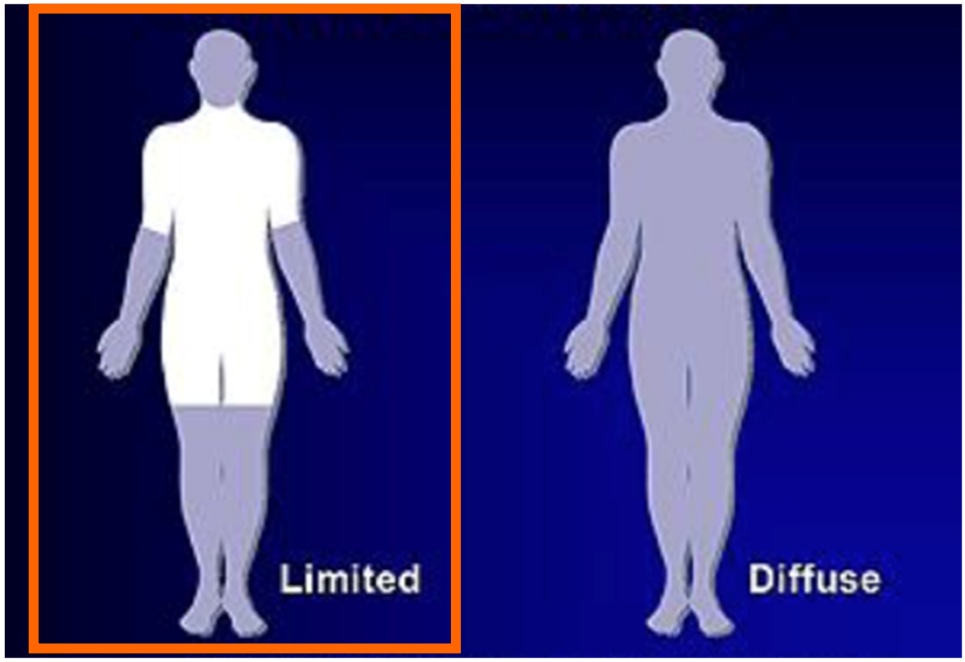
Limited Cutaneous

Skin
- Thickening occurs gradually
- Distal extremities, face, neck, upper chest.
- Telangiectasias and calcinosis common

GI
- Esophageal dysmotility > intestinal involvement

Pulm
- ILD in (17-35%)
- PAH more common and severe than diffuse disease

Renal
- Renal crisis uncommon



Anti-centromere (ACA) Ab common

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Pattanaik, et al. Front Immunol 2015; 6:272. Ostojic et al. Clin Rheum 25:453-57.
Medger T. in Clements and Furst 2nd Ed, Systemic Sclerosis

Diffuse Cutaneous SSc

Diffuse Cutaneous

Skin	• Thickening occurs early, more progressive • Extends to proximal extremities and trunk • Telangiectasias and calcinosis occur late • Tendon friction rub
GI	• Esophageal dysmotility • Intestinal disease more common
Pulm	• ILD (53-73)% • PAH less frequent than limited disease
Renal	• Renal Crisis more common

Anti-Topoisomerase 1 Ab (Scl-70)

Anti-RNA polymerase

Pattanaik, et al. Front Immunol 2015; 6:272. Ostojic et al. Clin Rheum 25:453-57.
Medger T. in Clements and Furst 2nd Ed, Systemic Sclerosis

Epidemiology and Mortality

- Prevalence:7.2-44.3 per 100,000
- Incidence 0.6-5.6 per 100,000
- Higher rates in:
  - US and Australia relative to Europe and Asia
  - African Americans
  - Females
- Ten year survival 65-73% in Europe and 54-82% in North America
- SSc-ILD in 70-80% of all SSc patients
  - 25-30% have progressive ILD

Bergamasco A., et al. Clin Epidemiol 2019;11:257-273
Khanna D., et al. Am J Resp Crit Care 2020 Med 201(6):650-660

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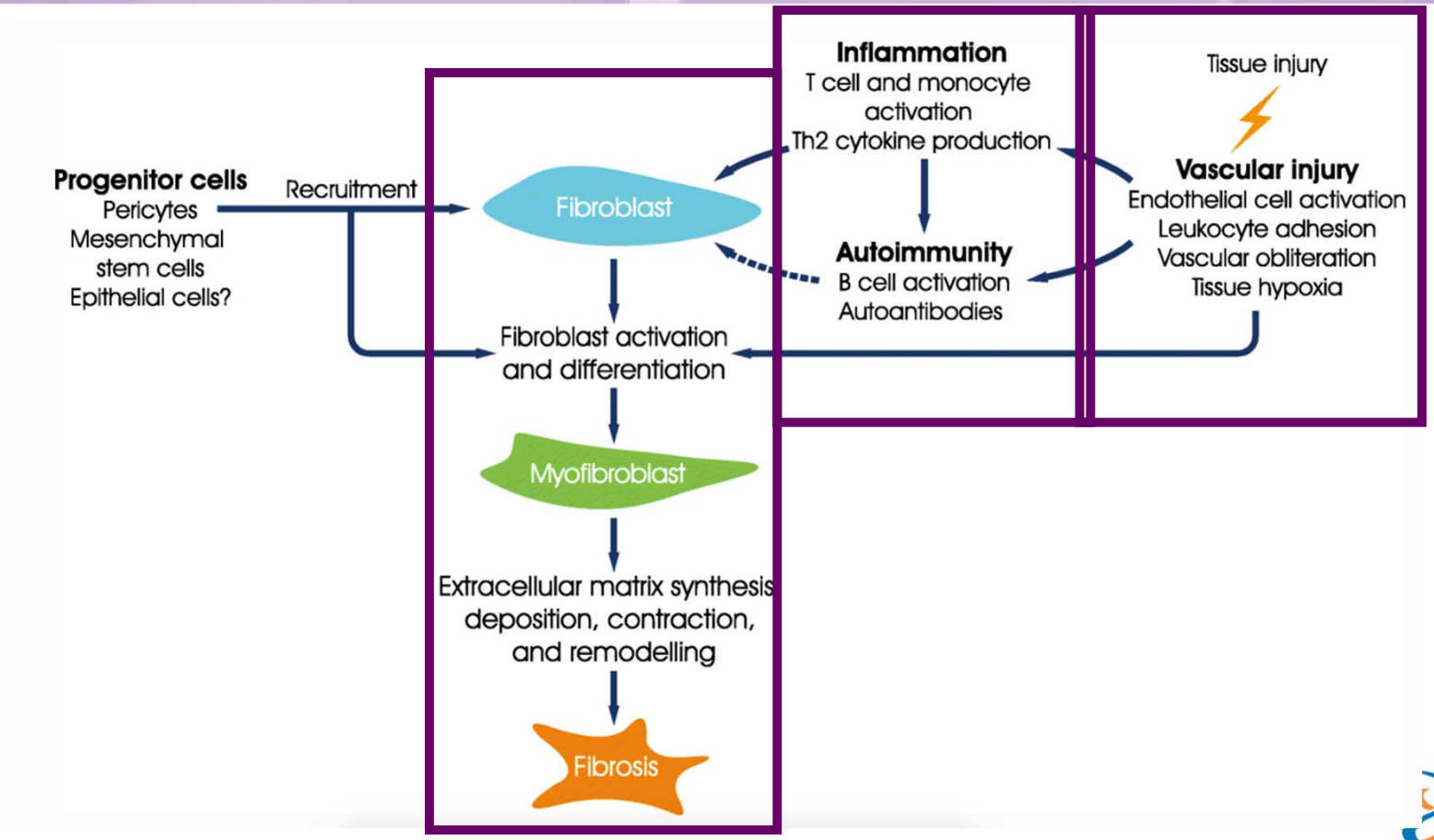
Pathogenesis



Varga and Abraham. Systemic Sclerosis: a Prototypeic Multisystem Fibrotic Disorder 2007; 117(3)
Cottin and Brown. Respiratory Research 2019: 20 :13

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Whiteboard Animation

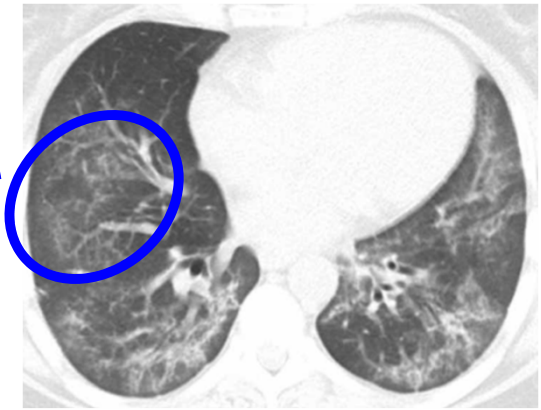
SSc-ILD Clinical Presentation

- ILD may occur *and may be severe* in both limited and diffuse cutaneous SSc
- < 5 years from the first scleroderma manifestation
- Scl-70 and ANA nucleolar pattern: both diffuse and limited disease
- Not all patients with SSc-ILD report respiratory symptoms
  - Cough, dyspnea
- PFTs: Restrictive pattern, but normal in some cases
  - Reduced FVC
  - Reduced DLCO may reflect ILD, PAH or both
- All patients should receive baseline PFTs, oxygen assessments, and HRCT on presentation.

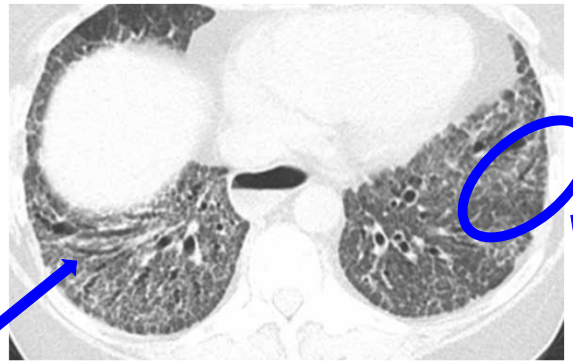
Clinical Presentation: HRCT

NSIP most common

- Basilar predominant
- Peribronchovascular
- Subpleural sparing
- Ground glass opacities
- Reticulation
- Traction bronchiectasis
- No honeycombing



NSIP Pattern
(Cellular with limited fibrosis)



NSIP Pattern
(Significant Fibrosis)

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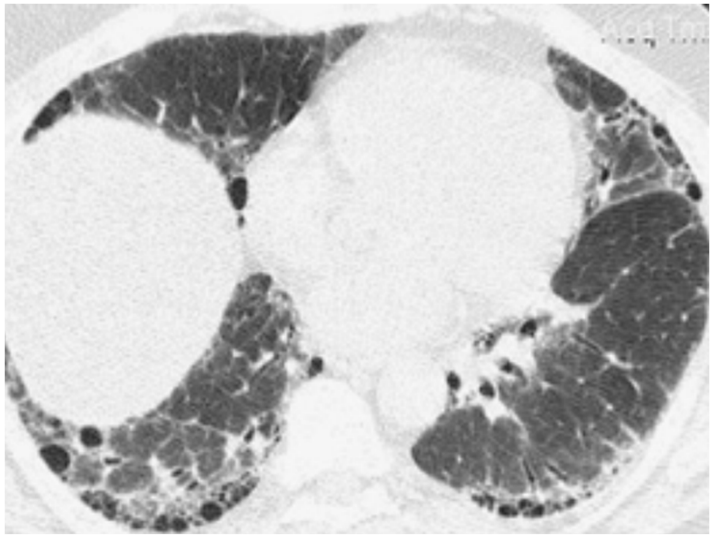
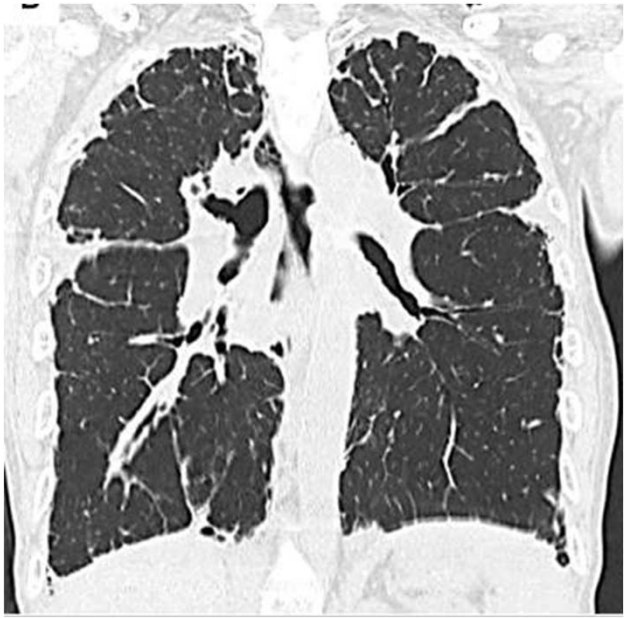
Clinical Presentation: HRCT



UIP Pattern



Pleuroparenchymal Fibroelastosis (PPFE)- poor prognosis

Radiologic *pattern* does <u>*not*</u> predict mortality

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Clinical Presentation: BAL

	UIP/ESL	NSIP	Cellular NSIP	Fibrotic NSIP
Subjects, n	10	57	12	45
Alveolar macrophages	82.5	78	76.5	79
	28–97	46–95	60–92	46–95
Lymphocytes	6	8	13.5	6
	1–22	0–45	6–30	0–45
Neutrophils	5	5	2.5	6
	1–55	1–41	1–12	1–41
Eosinophils	2.5	4	3	5
	0–4	0–19	0–10	0–19

No marked lymphocytosis

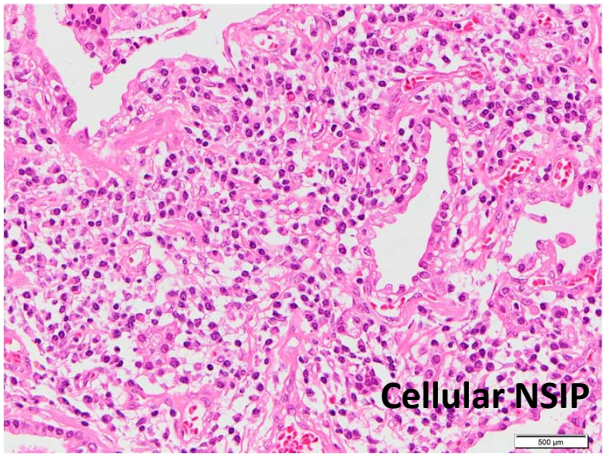
Bouros, et al. Am J Resp Crit Care Med. 2002;165:1581—1586.

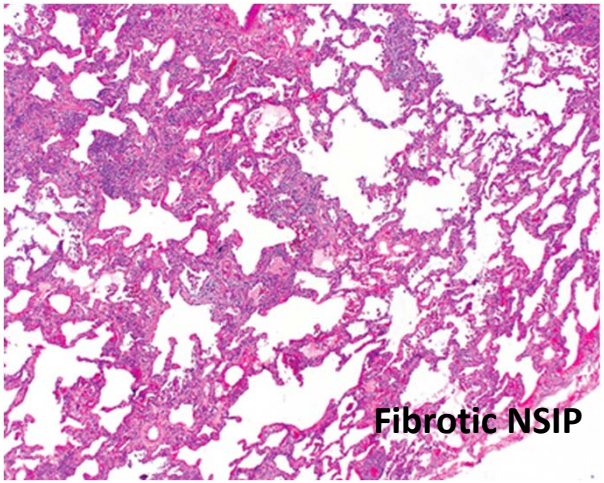
Clinical Presentation: Pathology

Histologic Subset	No. of Subjects	Type of Scleroderma (Limited/Diffuse)
NSIP	62 (77.5%)	43/19
UIP	6 (7.5%)	4/2
ESL	6 (7.5%)	5/1
Miscellaneous*	6 (7.5%)	4/2

80 SSc-ILD patients: NSIP in 78%, UIP 8%

Pattern did *not* predict mortality





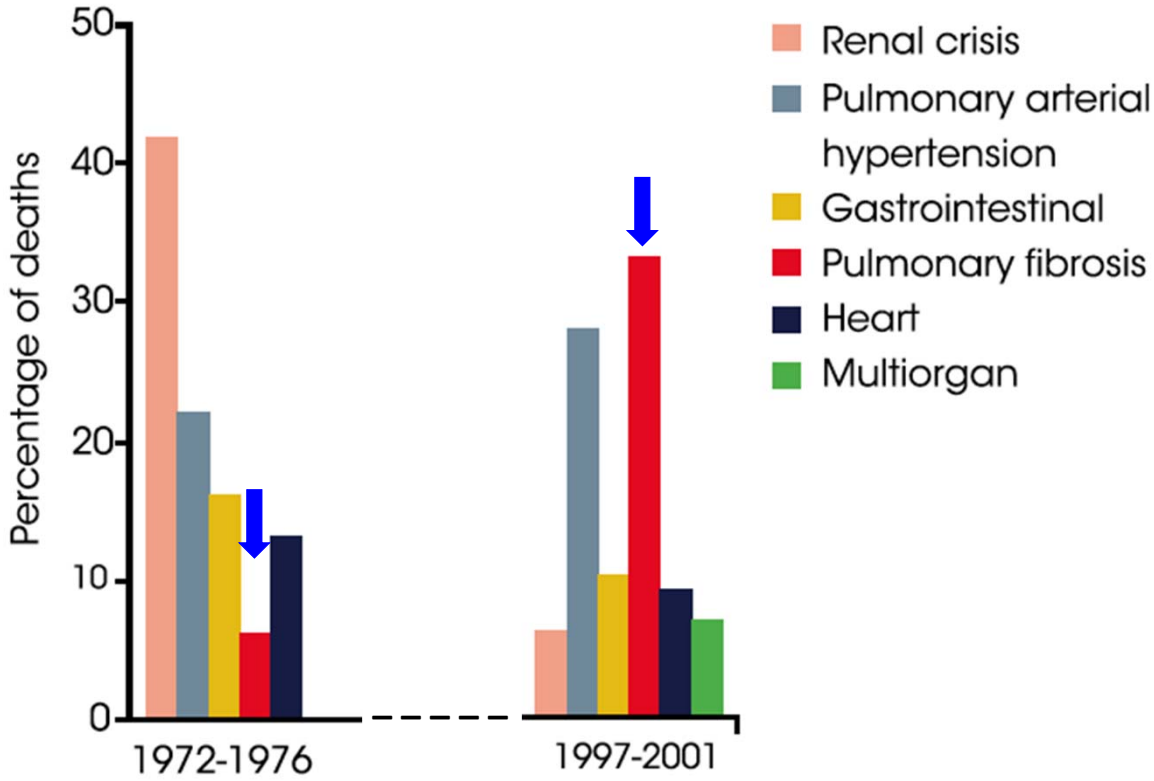
Bouros, et al. Am J Resp Crit Care Med. 2002;165:1581—1586.

Risk of Progression in SSc-ILD

Risk Factors for the Development or Progression of ILD

- Male gender, older age
- African American
- Diffuse Cutaneous
- Early disease – within 5 years of diagnosis
- Autoantibodies
  - Scl-70
  - Nucleolar pattern on ANA (anti-Th/To, anti-U3-RNP, anti-PM-Scl)
- Extent of disease on HRCT
- Decline (>10%) in FVC over 1 year

Pulmonary Fibrosis is now the leading cause of mortality in SSc



ALL patients should be screened with HRCT at the time of SSc diagnosis

35% of SSc-related deaths are due to pulmonary fibrosis

Steen VD, et al. Annals of the Rheumatic Diseases 2007;66:940-944.
Cottin and Brown. Respiratory Research 2019: 20 :13

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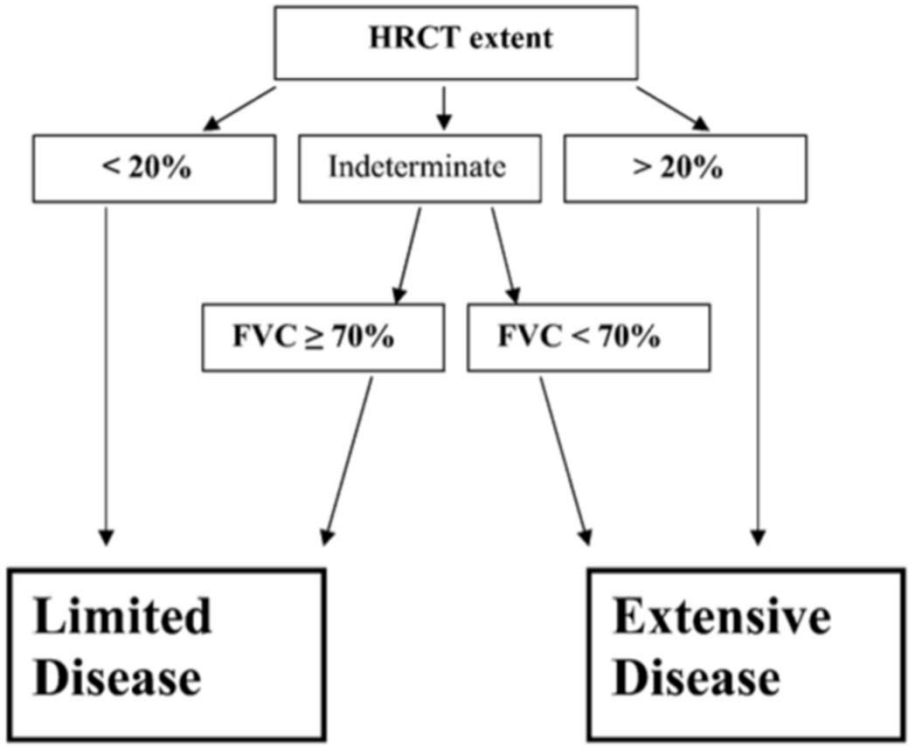
Predictors of Mortality: Staging SSc-ILD with HRCT and PFTs

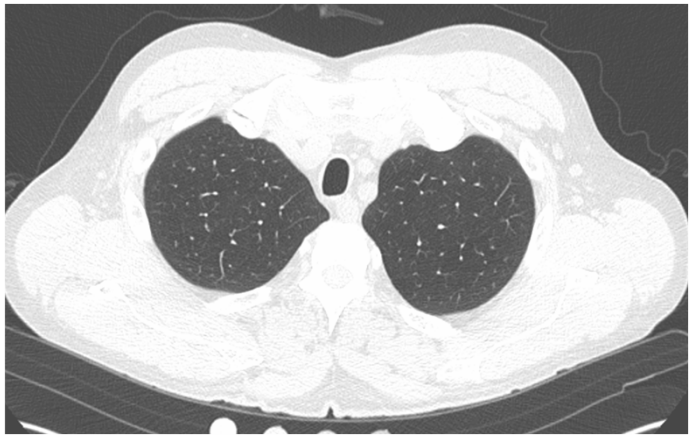
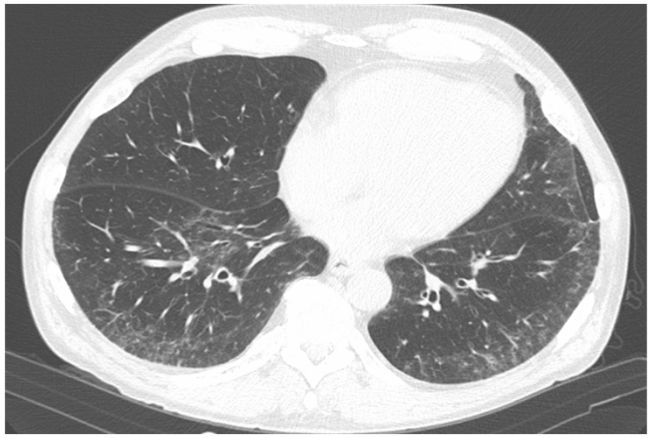
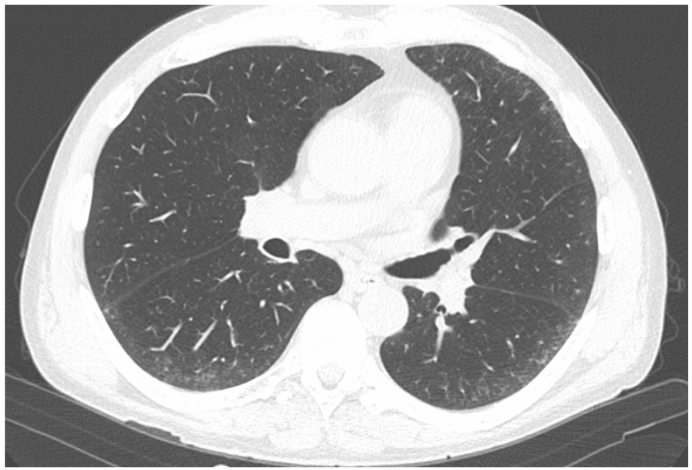
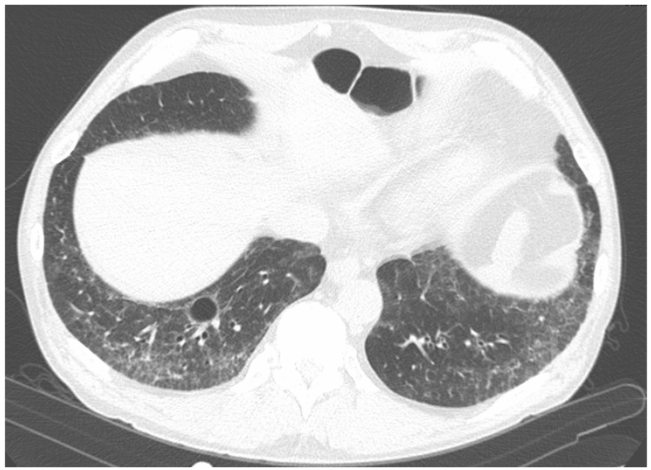


Limited vs Extensive disease predicts mortality

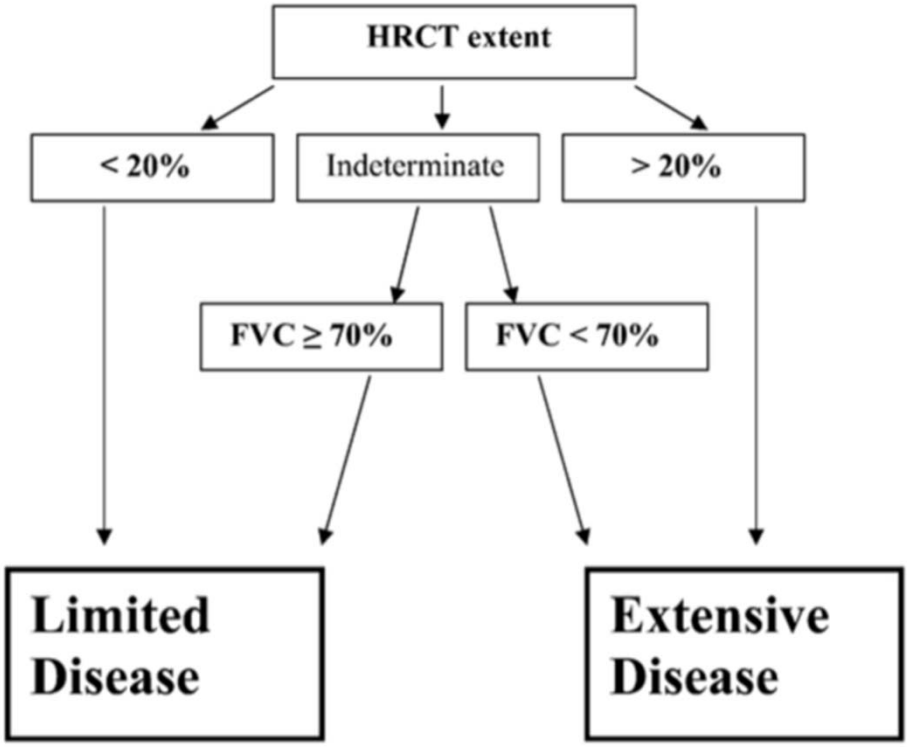
- Assess whether there is: <20% *or* >20% disease on HRCT
- If unclear, use FVC 70%

Goh NS et al, Am J Respir Crit Care Med 2008;177:1248-1254

Extent of Disease: Staging SSc-ILD with HRCT and PFTs

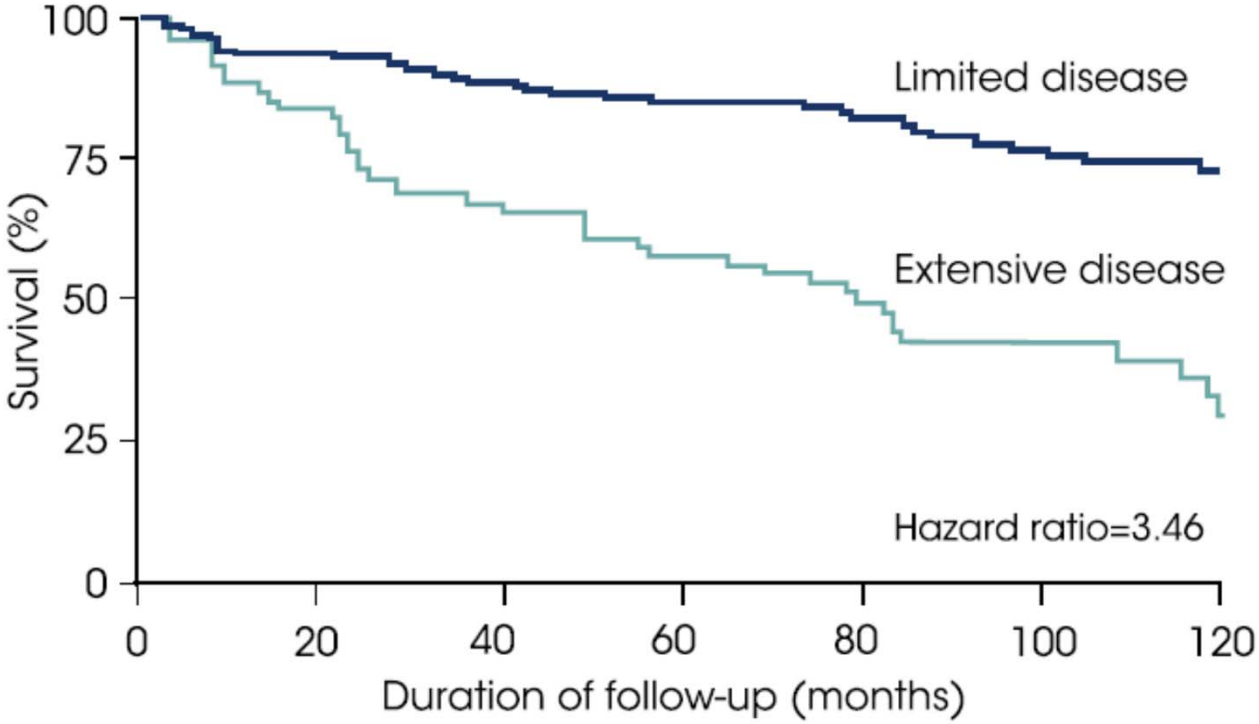





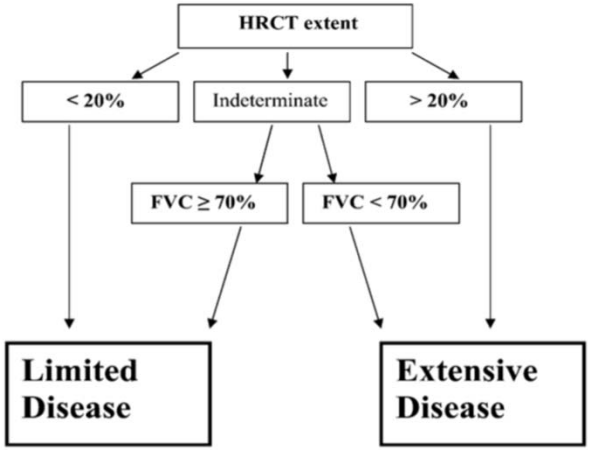
Extent of Disease: Staging SSc-ILD with HRCT and PFTs



FVC= 82% predicted

Predictors of Mortality: Staging SSc-ILD with HRCT and PFTs

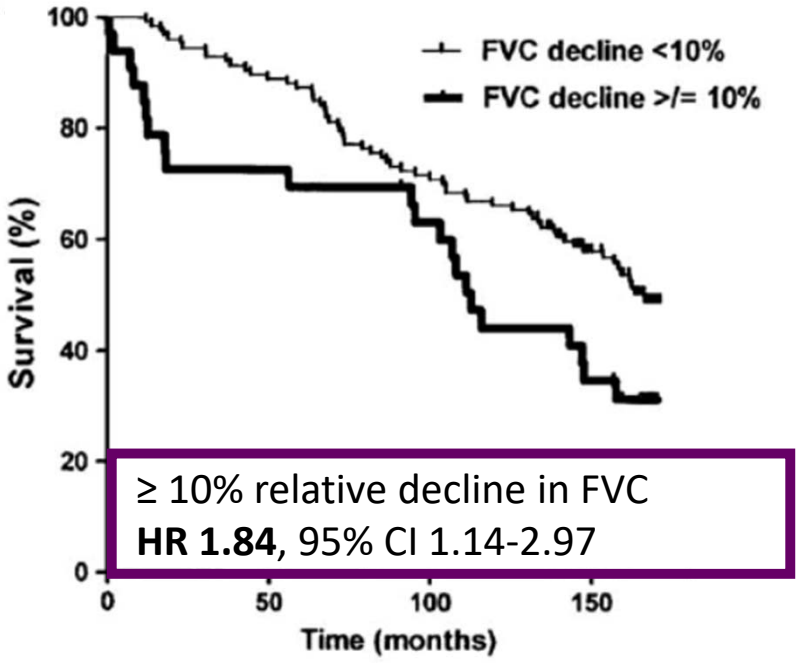




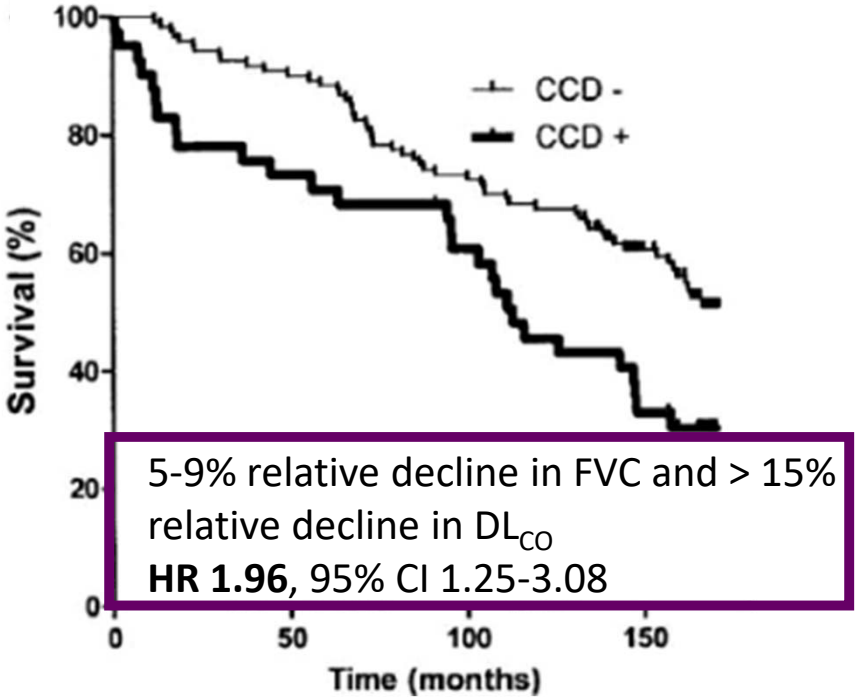
Goh NS, et al. Am J Respir Crit Care Med 2008;177:1248–54.
Cottin and Brown. Respiratory Research 2019: 20 :13

SSc-ILD Predictors of Mortality: PFTs

PFT trends at 1 year predict mortality



≥ 10% relative decline in FVC
HR 1.84, 95% CI 1.14-2.97



5-9% relative decline in FVC and > 15% relative decline in DL_{CO}
HR 1.96, 95% CI 1.25-3.08

Goh NS, et al. Arthritis Rheumatol 2017;69:1670-1678

SSc-ILD Disease Course

- ILD develops within 5 years of first non-Raynaud's symptom
- Most FVC decline occurs in the first 4 years after SSc diagnosis
- Variable disease course
  - Some have stable disease
  - Many experience slow progression
  - Some progress rapidly

Concerning Features for Pulmonologists

- Recent diagnosis
- Presence of Scl-70 Ab
- Absence of anti-centromere Ab
- Declining FVC (>10% of FVC per year)
  - Isolated Declining DLCO could be PAH
- Extent of Disease on HRCT

These should guide management decisions

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Chapter 2: Evidence Based Treatment Selection for SSc-ILD

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Professor
Chief, Division of Rheumatology
Director, Rheumatology Fellowship Program
Department of Medicine
Georgetown University
Washington, DC

Clinical features associated with limited and diffuse scleroderma

Limited cutaneous

Raynaud's -1st symptom alone for many years

General symptoms rare

Puffy FINGERs

Limited skin thickening

GI common, late PAH, and some lung fibrosis.

Diffuse cutaneous

Raynaud's often delayed

Acute onset, lots of constitutional symptoms

Arthralgias, carpal tunnel

Tendon friction rubs

Swollen, puffy HANDS

Early diffuse skin

Early organ involvement

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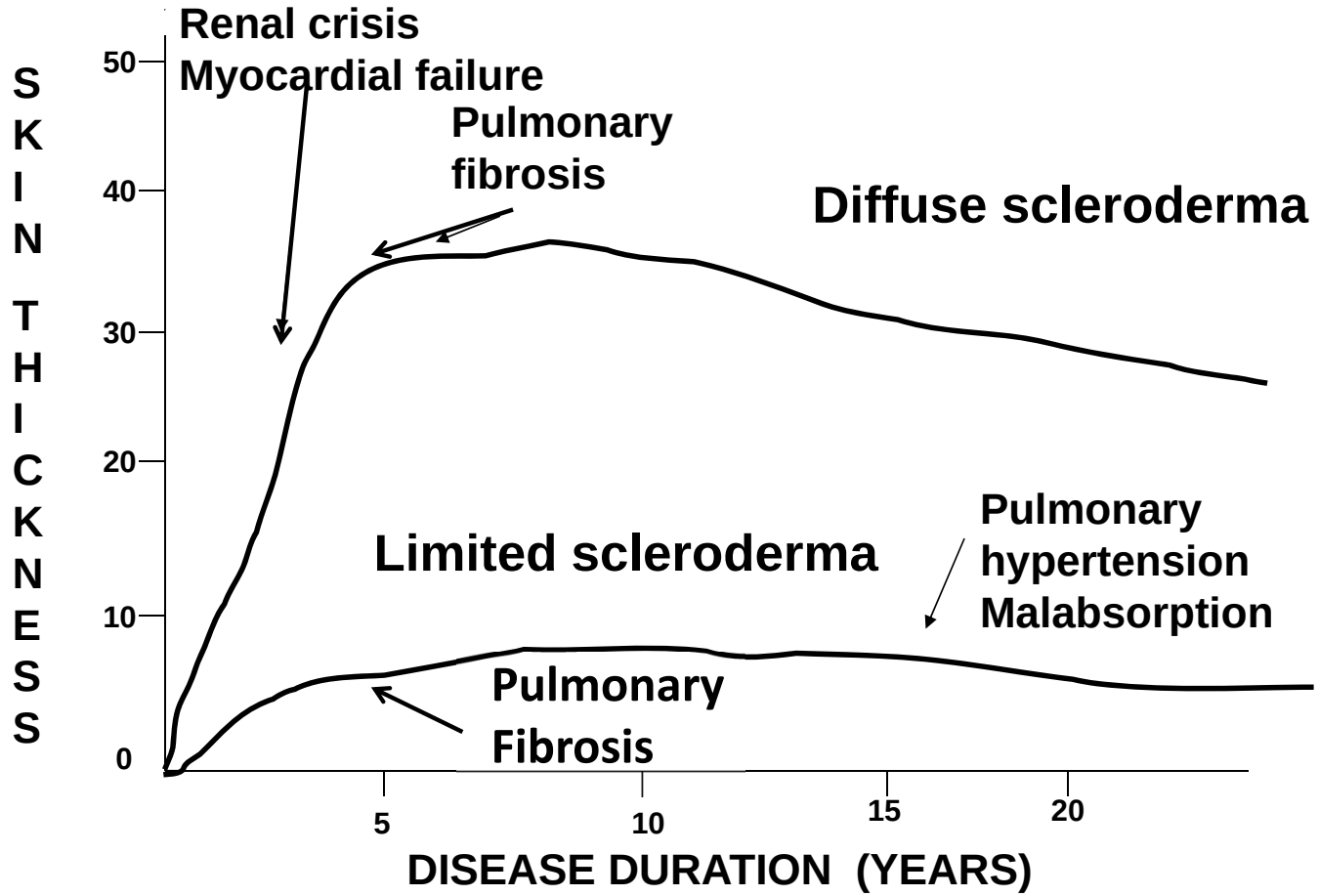
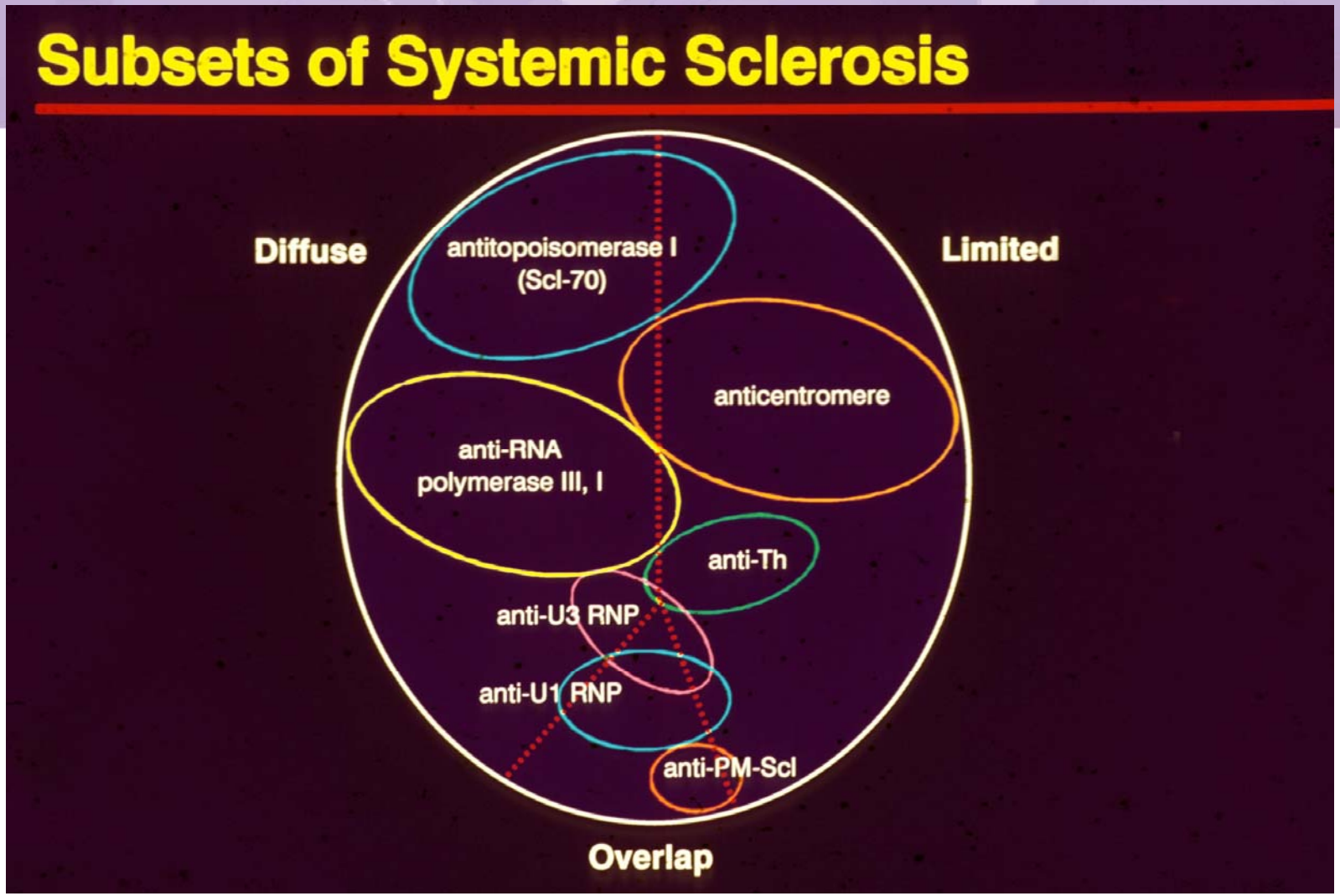
NATURAL HISTORY OF SCLERODERMA SUBSETS



Subsets of Systemic Sclerosis



Caveats in Management of Interstitial Lung Disease in SSc

- Fibrosis occurs early, but may stabilize later
- Use autoantibodies to help decide whether to treat.
- Having some fibrosis does not require treatment: extent and rapidity of change
- New SOB does not always mean active disease, and could mean something else.
- Steroids are not necessary for treatment.

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Features that do NOT suggest Fibrosis that needs treatment

- Anti-centromere antibody
- Isolated ↓ DLCO (in SSc)
- New, acute SOB with prior stable FVC < 65%: less likely active disease, MORE likely aspiration, CHF, PAH, pleural effusions, cancer
- New pleural effusions more likely cancer than fibrosis

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Treatment of SSc-ILD

Scleroderma Lung Study I

- Double blind, placebo controlled, cyclophosphamide vs placebo in patients.
- Cyclophosphamide group improved:
  - FVC difference of 2.94% predicted
  - NOT related to BAL or HRCT, only having significant fibrosis
  - HRCT better, but FVC fell after stopping treatment.
  - No survival benefit at 10 years.

Tashkin DP, Elashoff R, Clements PJ, et al. Cyclophosphamide versus placebo in scleroderma lung disease. *N Engl J Med*. 2006;354(25):2655-2666.

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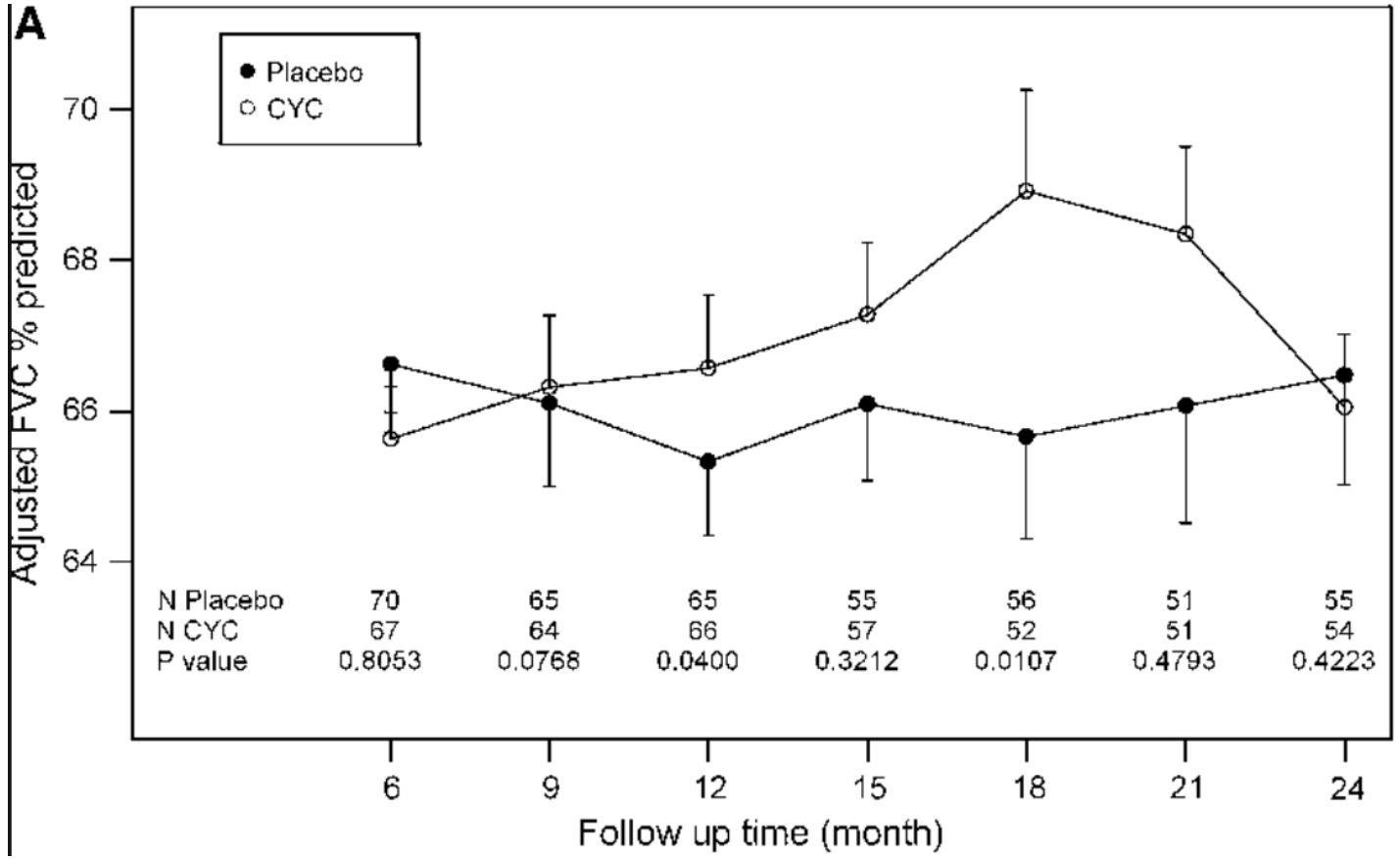
Scleroderma Lung Study I



Wells AU, Latsi P, McCune WJ. Daily cyclophosphamide for scleroderma: are patients with the most to gain underrepresented in this trial? Am J Respir Crit Care Med. 2007 Nov 15;176(10):952-3. doi: 10.1164/rccm.200708-1185ED. PMID: 17984310.

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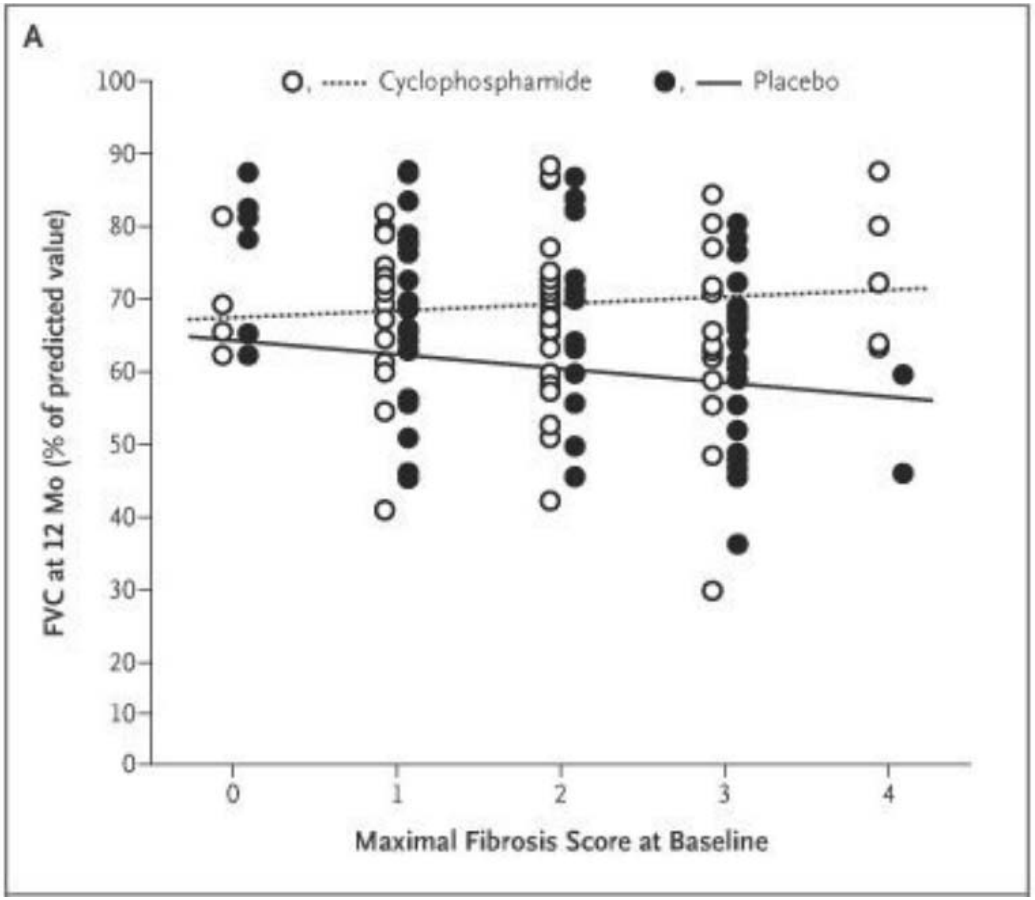
Improvement associated with more fibrosis



Tashkin DP, Elashoff R, Clements PJ, et al. Cyclophosphamide versus placebo in scleroderma lung disease. *N Engl J Med*. 2006;354(25):2655-2666.

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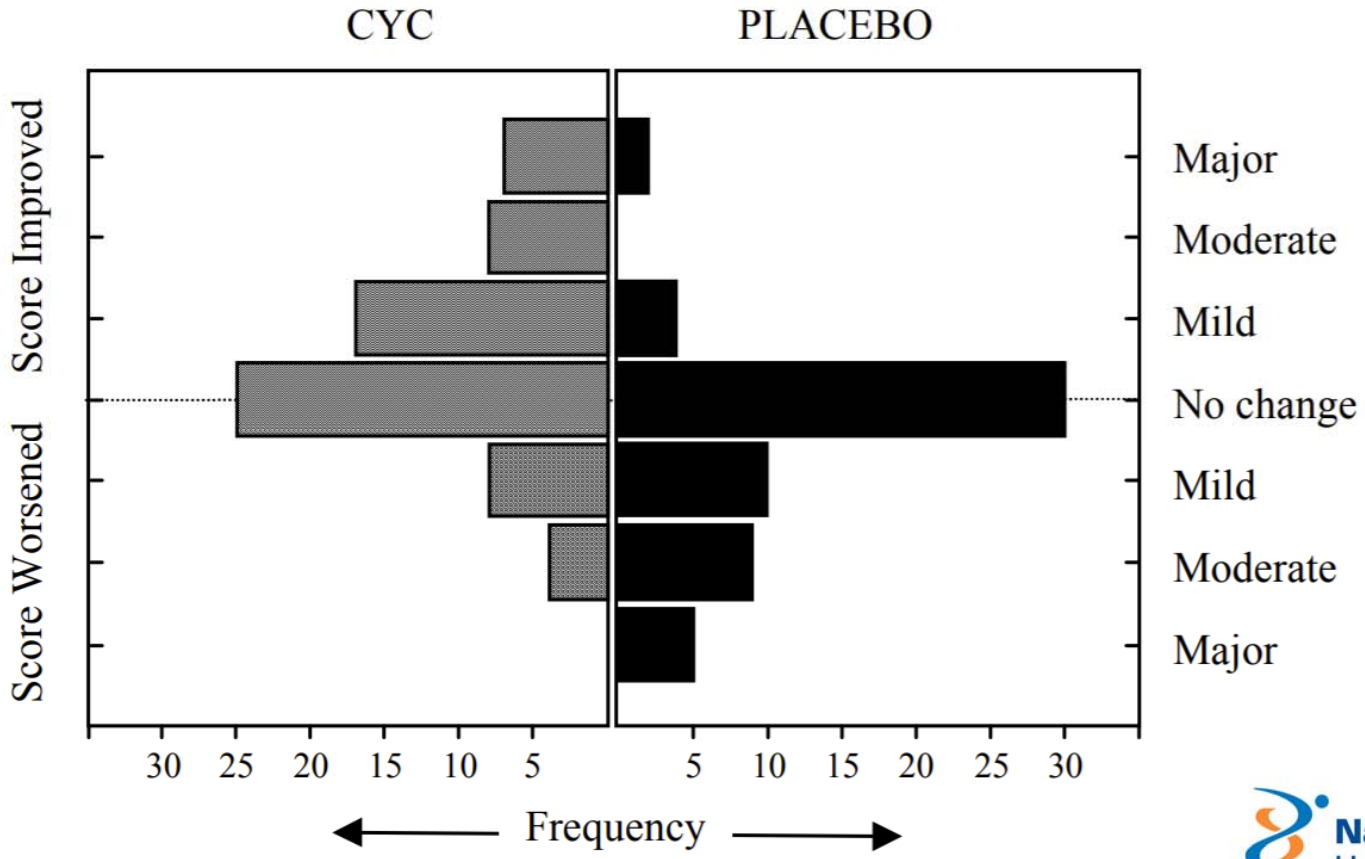
Scleroderma Lung Study I
Mahler (TDI) - Symptoms



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Tashkin DP, Elashoff R, Clements PJ, et al. Cyclophosphamide versus placebo in scleroderma lung disease. *N Engl J Med*. 2006;354(25):2655-2666.

Scleroderma Lung Study II

- Double blind, 1 year oral cyclophosphamide (+1 year placebo) vs 2 years mycophenolate mofeteil in patients with early (<7 years) SSc with ILD

- 142 patients: 52 years, 60% diffuse, mean 2.5 years disease duration; FVC 66%, DLCO 54%, 26% fibrosis on CT

Tashkin DP, Roth MD, Clements PJ, et al. Mycophenolate mofetil versus oral cyclophosphamide in scleroderma-related interstitial lung disease (SLS II): a randomised controlled, double-blind, parallel group trial. *Lancet Respir Med*. 2016;4(9):708-719. doi:10.1016/S2213-2600(16)30152-7

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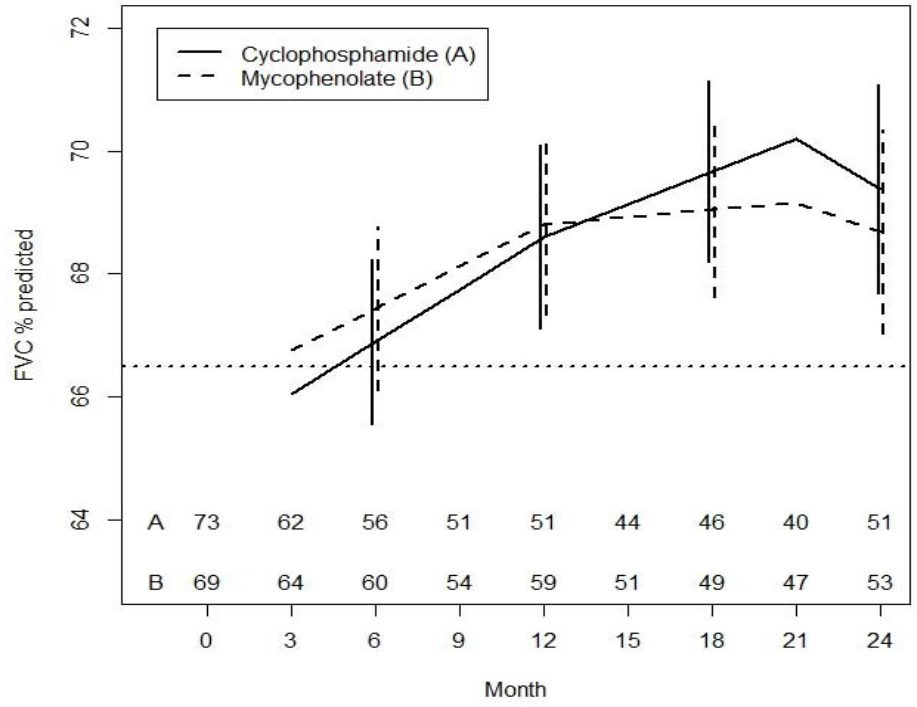
Scleroderma Lung Study II



cyclophosphamide 2mg/kg

MMF 3 gms/day

No difference between drugs,
Both improved

Tashkin DP, Roth MD, Clements PJ, et al. Mycophenolate mofetil versus oral cyclophosphamide in scleroderma-related interstitial lung disease (SLS II): a randomised controlled, double-blind, parallel group trial. *Lancet Respir Med*. 2016;4(9):708-719. doi:10.1016/S2213-2600(16)30152-7

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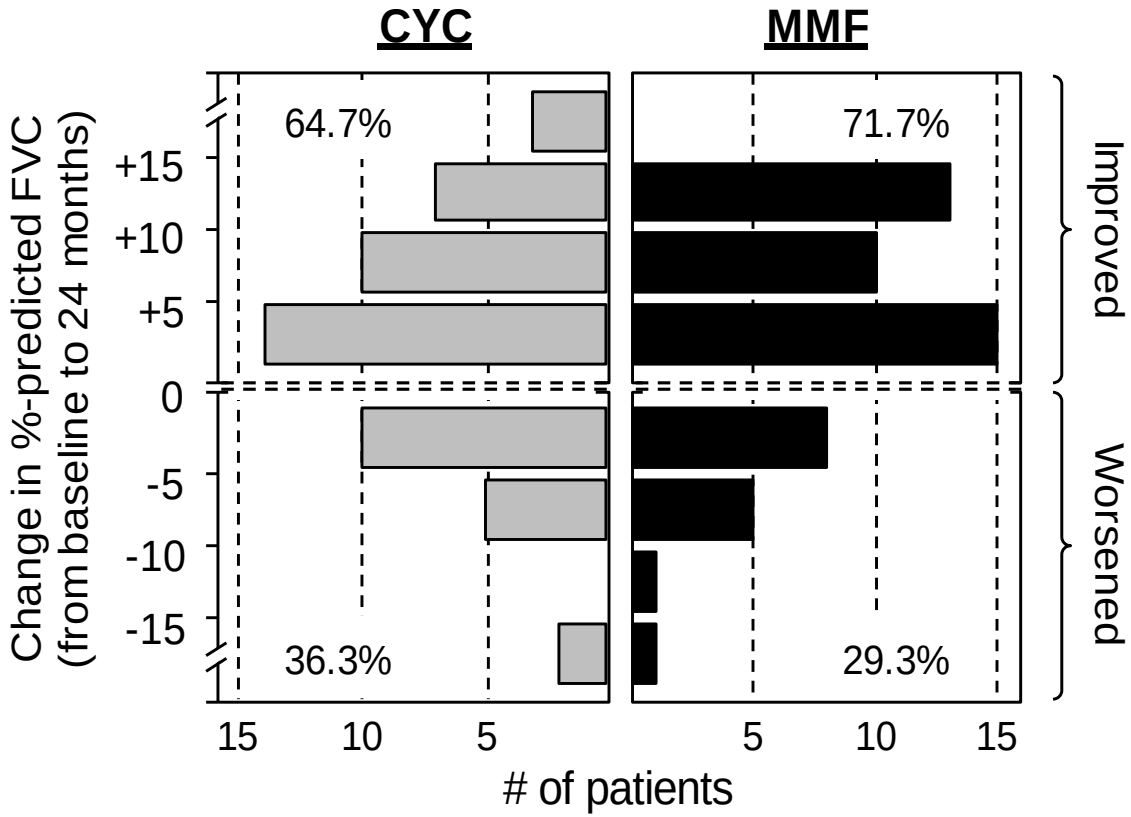

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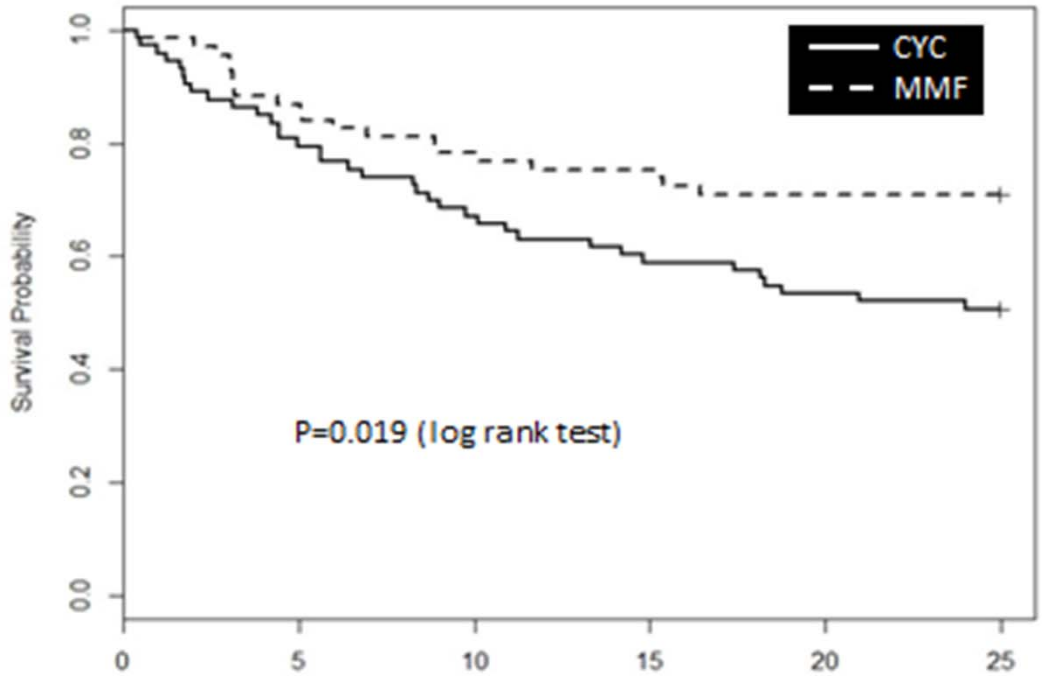
Scleroderma Lung Study II

Time to withdrawal



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Treatment in Early Disease

- Guidelines (and standard of care) say that immunosuppressive agents should be used early in SSc – ILD.
- Improves lung function, including symptoms, as well as skin, and well being.
- Mycophenolate mofetil is better tolerated than cyclophosphamide with improvement in lung function

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Immunosuppressive Therapy

- In some patients, there is the ultimate development of progressive lung fibrosis – despite immunosuppressive therapy.

Antifibrotics: Nintedanib and Pirfenidone

In 2014, the FDA approved 2 agents for Idiopathic Pulmonary Fibrosis (IPF) - which has a Usual Interstitial Pneumonia (UIP) pattern of fibrosis.
Both slowed down rate of FVC decline by ~ 50% over 1 year.

Nintedanib
- Triple tyrosine kinase inhibitor: PDGF, VEGF, FGF receptors
- Most common side effect: Diarrhea (62%)
- Dose: 150mg po BID
- Monitor LFTs

Pirfenidone
- Antioxidant, anti-inflammatory, and anti-fibrotic properties
- Most common side effects: Nausea, weight loss, photosensitivity rash
- Dose: 801mg po TID
- Monitor LFTs

Richeldi L , et al. N Engl J Med. 2014 ;370:2071-2082.
King TE, et al. N Engl J Med. 2014;370:2083-2092.

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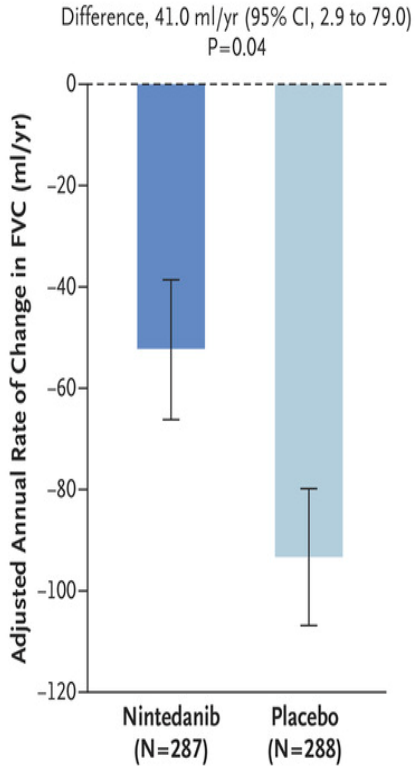
Nintedanib in Systemic Sclerosis SENSCIS

SENSCIS Trial (NEJM 2019)

- 576 patients with SSc randomized to nintedanib or placebo for one year
  - HRCT with fibrosis >10% lungs
  - Disease onset within 7 years
  - FVC > 40%
  - DLCO 30-89%
- Background therapy with MMF (48%), MTX (6.6%), or prednisone < 10mg



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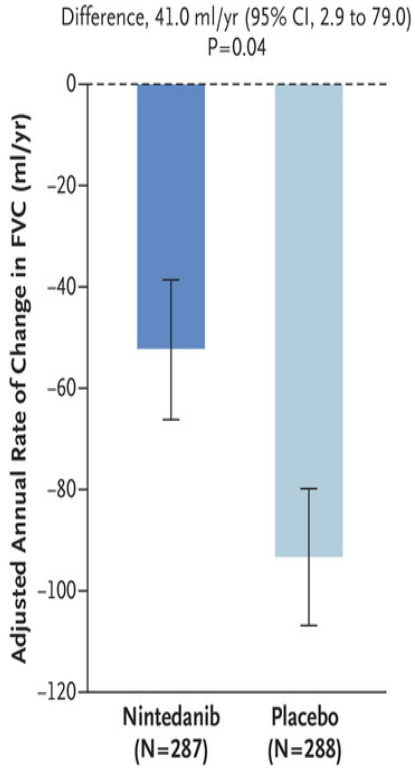
Distler et al, NEJM 2019; 2518-2528.

Nintedanib in Systemic Sclerosis SENSCIS

SENSCIS Trial (NEJM 2019)

- Primary outcome: Adjusted annual rate of decline of FVC (mL/year)
- No differences in symptoms or other clinical signs of SSc.
- Adverse effects included diarrhea (76% vs 32%) and LFT elevations (4.9 vs 0.7%)
- Nintedanib discontinued in (16% vs 8.7%)
- Placebo lost 93.3 mL/yr vs Nintedanib lost 52.4 mL/yr = Δ 41.0 ml/year



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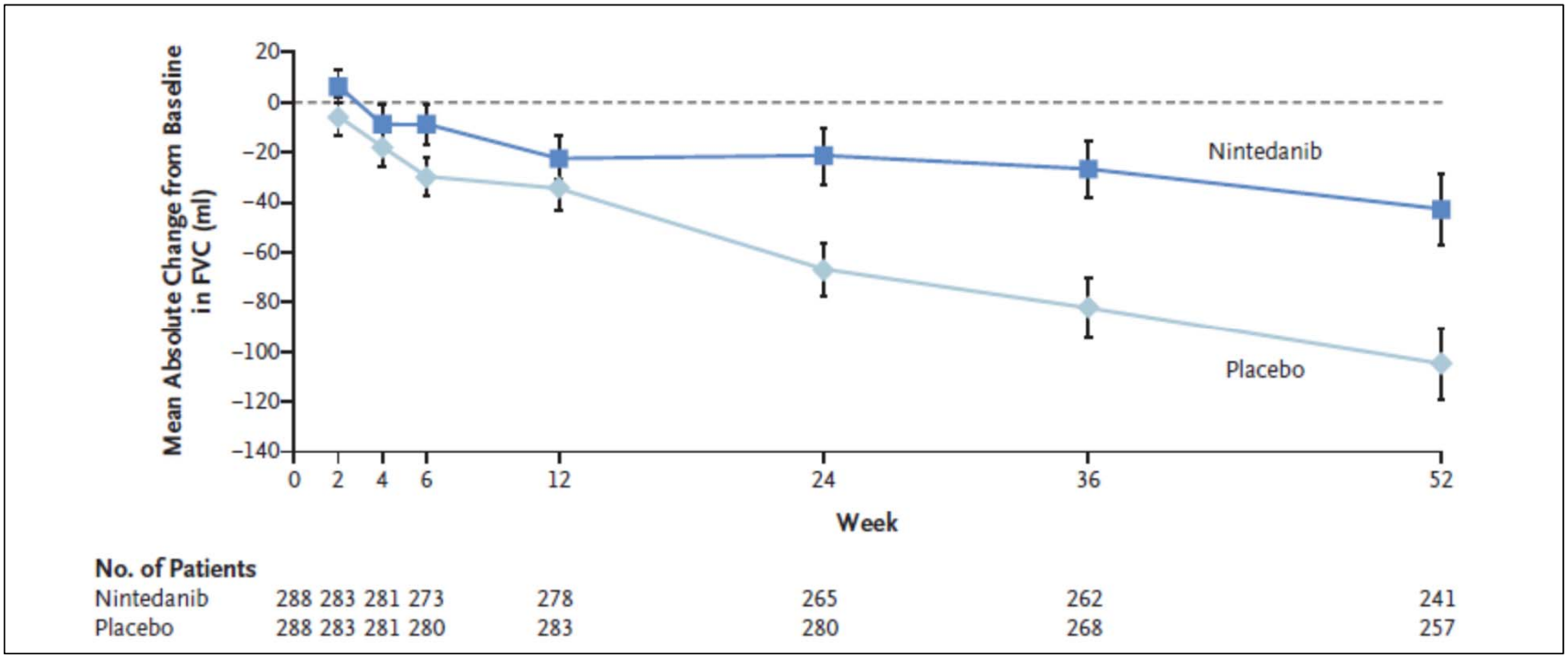
Distler et al, NEJM 2019; 2518-2528.

Nintedanib in Systemic Sclerosis SENSCIS



Distler et al, NEJM 2019; 2518-2528.

Nintedanib in Systemic Sclerosis

- Effective therapies to treat interstitial lung disease in SSC (CYC, MMF) and in this study 48% received background therapy with MMF

	Nintedanib	Placebo	
All patients	-52.4 mL/yr	-93.3 mL/yr	41mL/yr (95% CI 3-79mL)
Background MMF	-40.2 mL/yr	-66.5 mL/yr	Less of a benefit?
No MMF therapy	-63.9 mL/yr	-119.3 mL/yr	More of a benefit?

Distler et al, NEJM 2019; 2518-2528.

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Nintedanib in Systemic Sclerosis

- However, the authors cautioned against over interpreting multiple subgroup analysis as a means of identifying independent predictors of treatment response.

	Nintedanib	Placebo	Change
All patients	-52.4 mL/yr	-93.3 mL/yr	41mL/yr (95% CI 3-79mL)
Background MMF	-40.2 mL/yr	-66.5 mL/yr	Less of a benefit?
No MMF therapy	-63.9 mL/yr	-119.3 mL/yr	More of a benefit?

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Distler et al, NEJM 2019; 2518-2528.

Nintedanib in Systemic Sclerosis INBUILD

INBUILD trial

- 663 patients with progressive fibrosing lung disease (PF-ILD) other than IPF randomized to nintedanib or placebo
- FVC >45%
- DLCO 30-80%
- HRCT with fibrosis >10% lungs; stratified by UIP pattern
- Progression within the past 2 years
  - A relative decline of 10% predicted value
  - A relative decline of 5% to 10 % predicted value &
    - Increased symptoms or Increased extent of fibrosis

Flaherty KR, et al, NEJM, 2019.

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Nintedanib in Systemic Sclerosis INBUILD

INBUILD trial

- Primary outcome: Annual rate of decline of FVC (mL/year)
- No background therapy allowed initially—could be added if further progression at 6 months
- 25.6% had CTD-ILD; (52% had RA; 23% SSc)

Flaherty KR, et al, NEJM, 2019.

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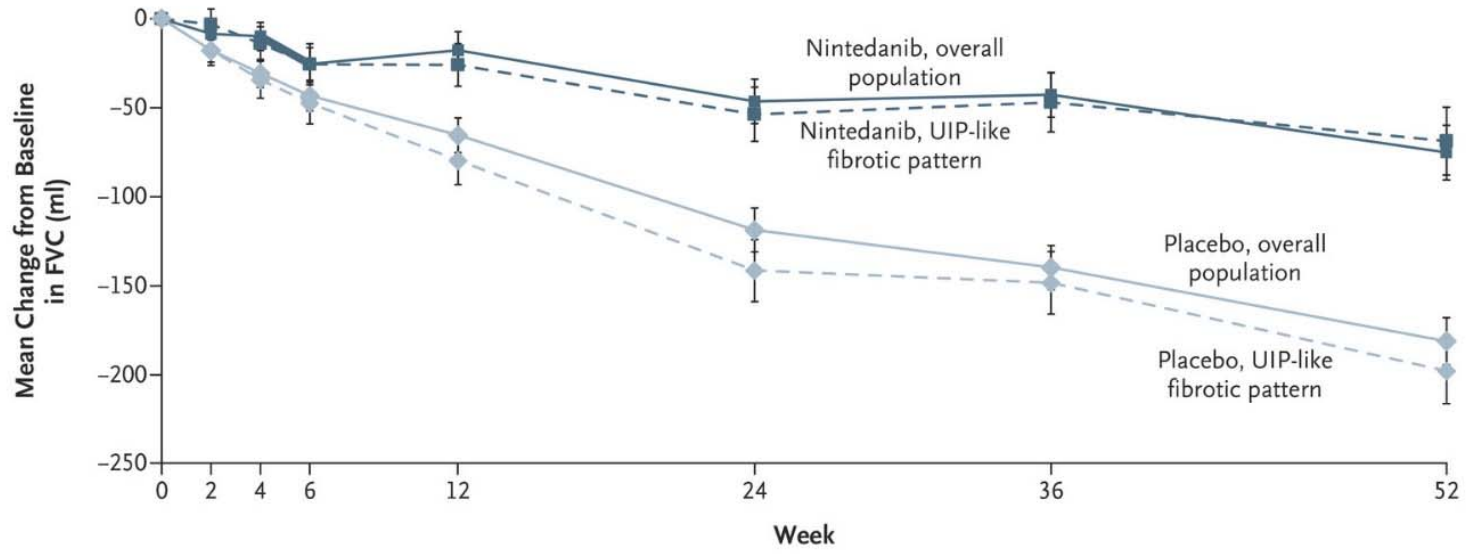
Nintedanib in Systemic Sclerosis INBUILD



No. of Patients								
Overall population								
Nintedanib	332	326	320	322	314	298	285	265
Placebo	331	325	326	325	320	311	296	274
Patients with UIP-like fibrotic pattern								
Nintedanib	206	203	200	199	193	180	171	160
Placebo	206	202	202	201	197	190	176	162

The adjusted rate of decline with Nintedanib = -80.8 mL/yr
Vs.
Placebo = -187.8mL/yr

The adjusted rate of decline with a UIP patter
Nintedanib = -82.9 mL/yr
Vs.
Placebo = -211.1 mL/yr

Flaherty KR, et al, NEJM, 2019.

Pirfenidone in Systemic Sclerosis

An Open-label, Phase II Study of the Safety and Tolerability of Pirfenidone in Patients with Scleroderma-associated Interstitial Lung Disease: the LOTUSS Trial

Dinesh Khanna, Carlo Albera, Aryeh Fischer, Nader Khalidi, Ganesh Raghu, Lorinda Chung, Dan Chen, Elena Schiopu, Margit Tagliaferri, James R. Seibold, and Eduard Gorina

Khanna D, et al. J Rheumatol 2016;43:1672.

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Pirfenidone in Systemic Sclerosis

- 63 patients, 18 sites, 3 countries
- All subjects received pirfenidone
  - 16 weeks total
  - 2403 mg/day
  - Randomized to 2 versus 4 week titration schedules

The Journal of Rheumatology 2016; 43:9; 1672-1679

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Pirfenidone in Systemic Sclerosis

- Most common treatment emergent adverse event (TEAE): nausea, headache, fatigue
- MMF use in 63.5%, did not affect tolerability
- 89% completed study
  - 6 withdrew due to TEAE, 5 in 2 week titration group and 1 in 4 week group
- TEAE occurred more often during titration than maintenance
- Severe TEAE was seen in 19%, mostly occurred at full dose
- No change in lung physiology, dyspnea, skin thickness, HAQ-DI or ptGA score at 16 weeks

Conclusion: similar tolerability profile as IPF trials; well tolerated with MMF

The Journal of Rheumatology 2016; 43:9; 1672-1679

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Pirfenidone in Systemic Sclerosis Ongoing Trials

- Scleroderma Lung Study (SLS) III
  - Phase II RCT, PFD + MMF vs. Plac + MMF; goal n = 150 patients
  - Primary endpoint: Change in FVC % predicted over 18 months
  - Secondary endpoints: Change in mRSS, extent/total fibrosis on HRCT, etc.
  - PI: Roth (UCLA), Genentech (NCT03221257)

www.clinicaltrials.gov; Last Accessed 10/12/2020.

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Initial Evaluation

- In summary …
  - "Not all rules are absolute…"
  - ILD should be suspected in anyone diagnosed with SSc
  - Initial evaluation should include:
    - Assessment of Respiratory Symptoms
    - Clinical examination (Crackles)
    - Pulmonary Function Testing
    - High Resolution Computed Tomography (HRCT)
    - Gas exchange (Hypoxemia)
    - Screening for Pulmonary Hypertension

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Initial Therapy

- In summary …
  - As Dr. Steen said, consider early immunosuppressive therapy in those with:
    - High risk of progression
    - Present with clinically significant disease
  - Consider antifibrotic (nintedanib) therapy in those who show evidence of progression despite immunosuppressive therapy.

Chapter 3: Longitudinal Management of SSc-ILD

Amy Olson, MD, MSPH
Associate Professor
Department of Medicine
Division of Pulmonary,
Critical Care & Sleep Medicine
National Jewish Health
Denver, CO

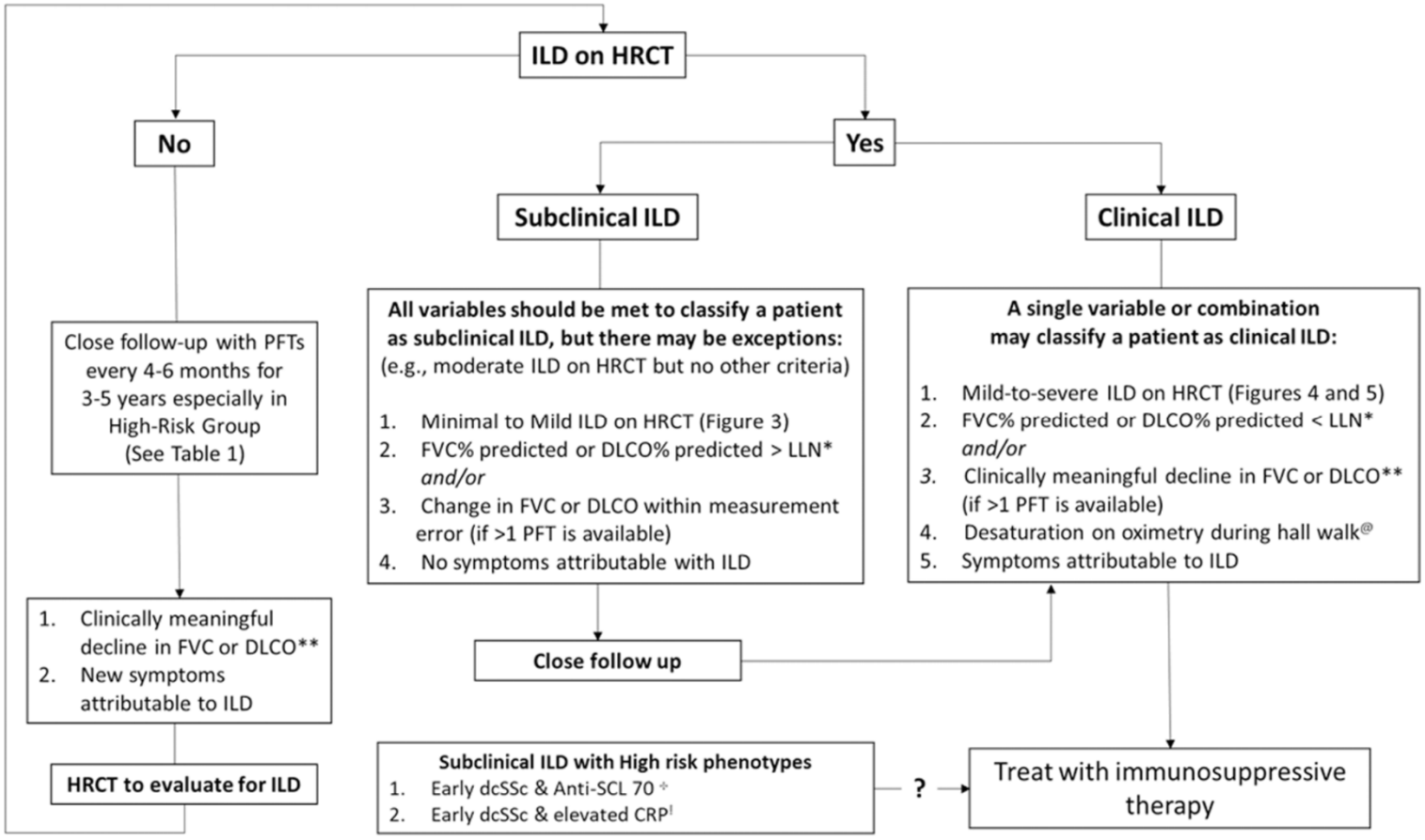
Longitudinal Monitoring:

"Universal screening is paramount in identifying patients early."

- 50% General Rheumatologists Screen
- 2/3 SSC Specialists Screen
- So let's screen and come up with our longitudinal monitoring plan…

Roofeh D, et al. Curr Opin Rheumatol 2019;31:241.

Longitudinal Monitoring:



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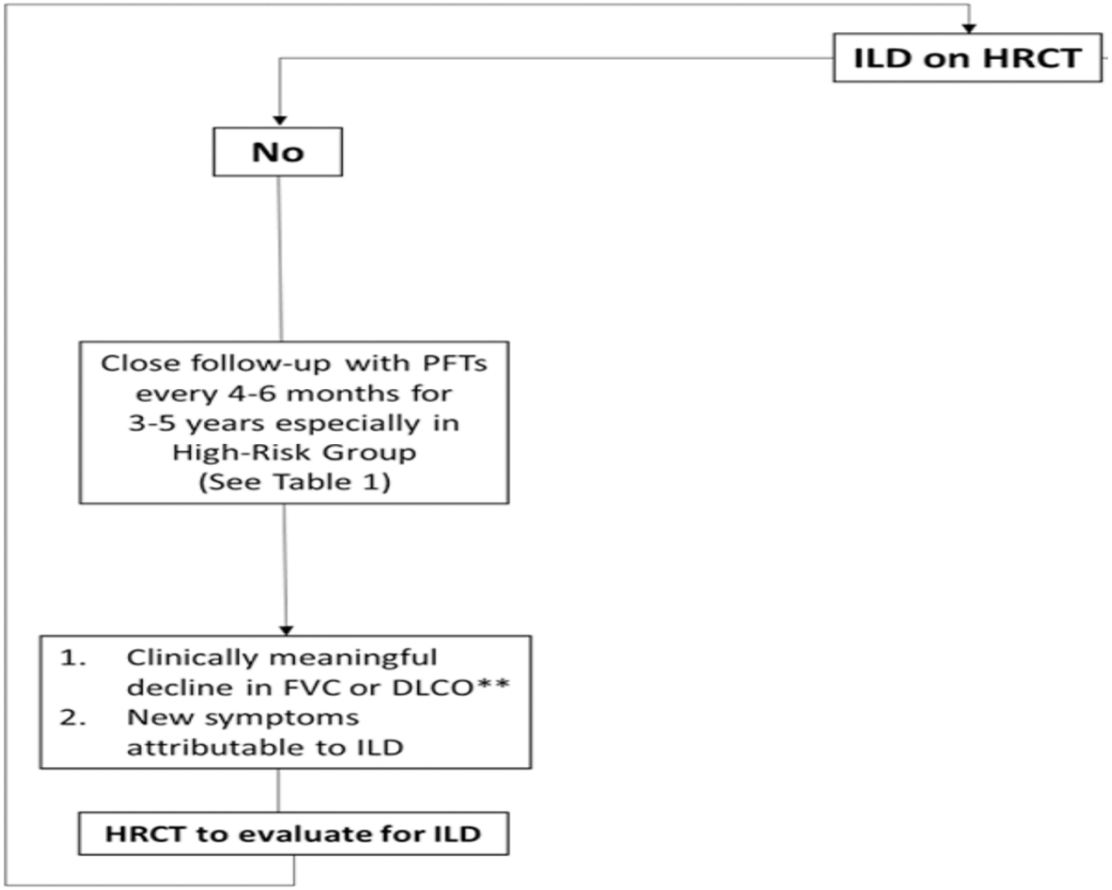

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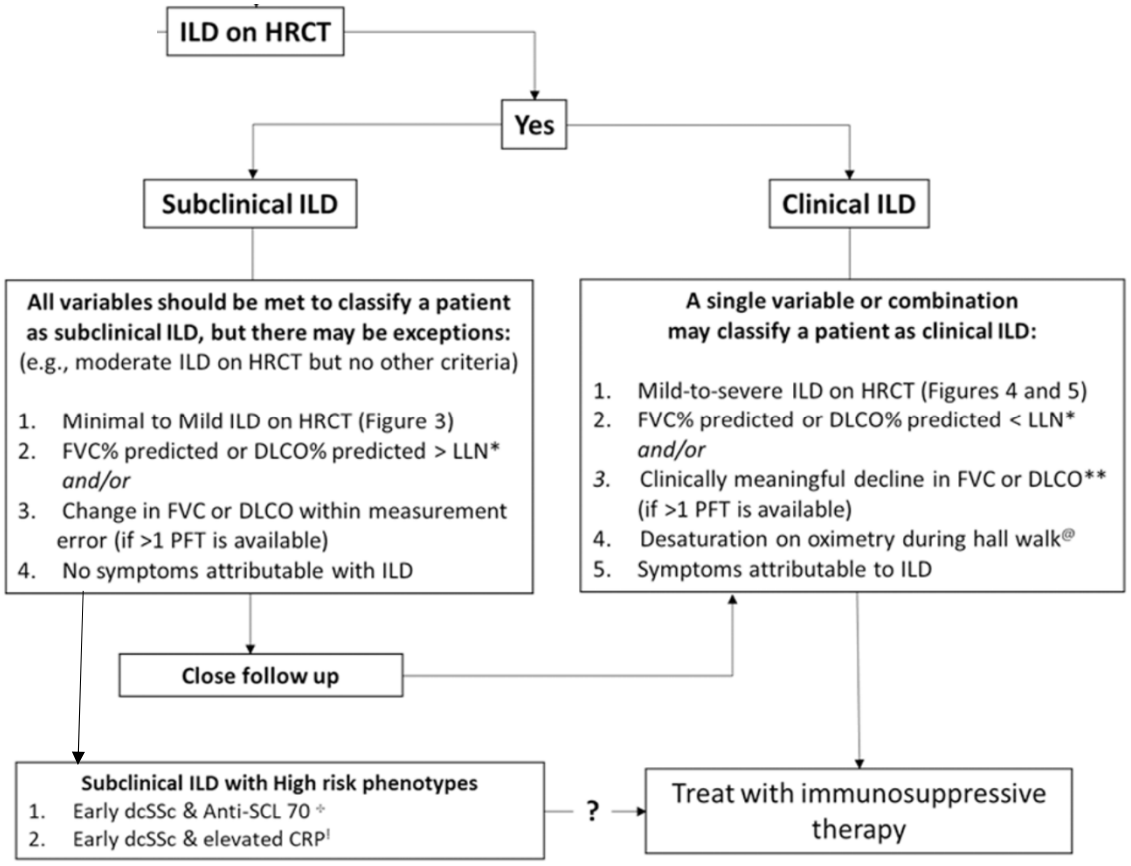

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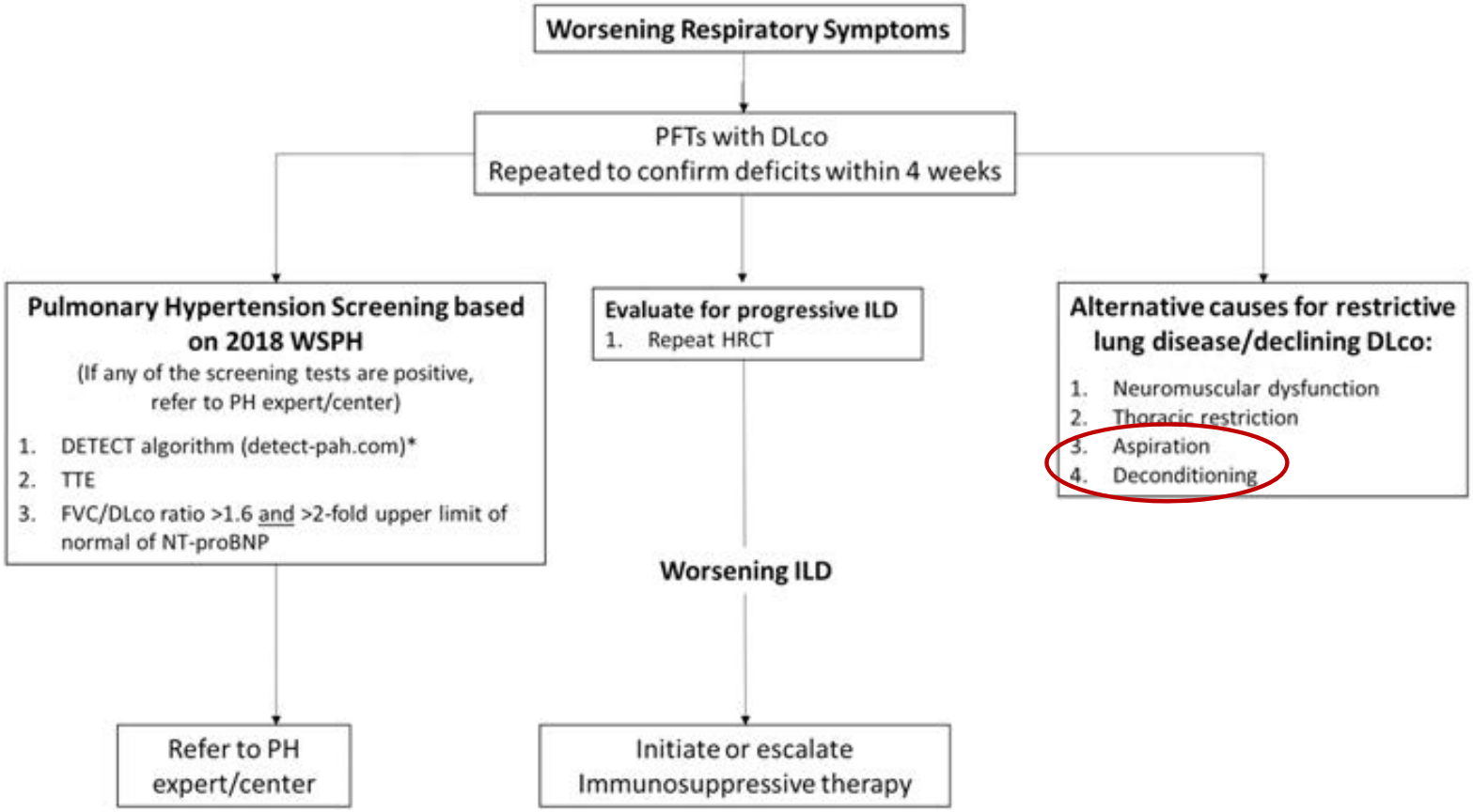

Roofeh D, et al. Curr Opin Rheumatol 2019;31:241.

Longitudinal Monitoring Summary:

- Monitoring:
  - Clinical Examination
    - Assess for worsening breathlessness or cough and causes (esophageal dysmotility*)
    - Assess for extra-pulmonary SSC symptoms
  - Pulmonary function testing
    - Declines
  - Oxygen titration (forehead/ear probe) & 6-minute walk distance
  - HRCT and CT imaging
    - Change in symptoms
    - Annually*?
  - Echocardiogram/Right Heart Catheterization
    - Baseline and consider as the cause for worsening symptoms
  - Ensure laboratory monitoring for patients on pharmacotherapies
- How Often:
  - Clinically monitor at ~ 3 months (with PFTs) for the first ~ 3 to 5 years, or in those patients with evidence of progression.
  - See any patient sooner for any worsening pulmonary symptoms …

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A Multidisciplinary Team Provides An Overall Approach to Care

The Manifestation of Disease	The Team Member
Primary Care Physician	Coordination of Care
Systemic Sclerosis	Rheumatologist
SSc-ILD	Pulmonologist – ILD Radiology/Pathology
Pulmonary Hypertension	Pulmonologist/Cardiologist - pHTN
GERD & Esophageal Dysmotility	Gastroenterologist
Renal Disease/Renal Crisis	Nephrologist
Cardiac Disease	Cardiologist
Neuromuscular Disease	Neurology
Other Complications	A number of practitioners (ID, etc.)

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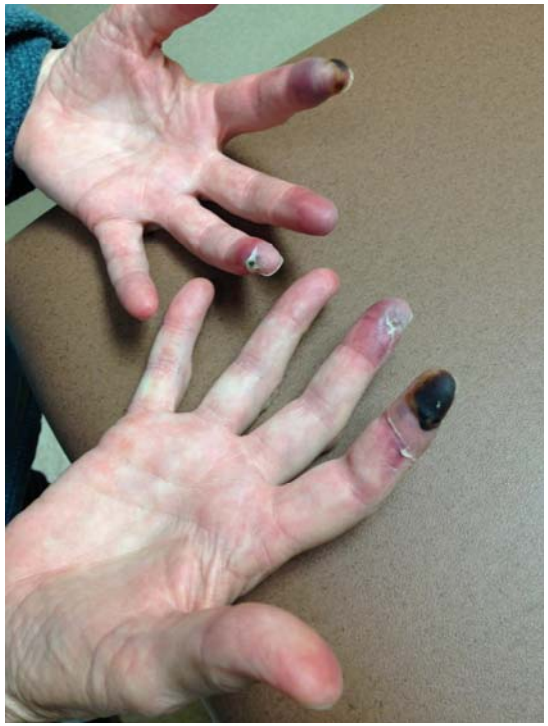

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Non-Pharmacologic Therapies

- Education about Disease
  - Comorbidities
- Stop Smoking
- Appropriate Nutrition
- Determine Need for Supplemental Oxygen
  - At rest, with ambulation, during exercise and sleep
- Supportive Care
  - For Patients & Caregivers
  - Support Groups
  - The Scleroderma Foundation
  - Palliative Care

- Nursing Support
  - Assess for new or worsening symptoms
  - Therapeutic Monitoring
- Pulmonary Rehabilitation
- Vaccinations/Ongoing Health Maintenance
- Lung Transplantation if Required
  - Observational studies are encouraging
- Clinical Trials!

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Patient Perspective Video

Patient Centered Communication:

Patient Priorities

- Minimize Uncertainty
  - Understand Disease
  - Understand Treatments
- Seek Assistance from Partner, Family, & Friends … BUT Maintain Independence
- Maintain Energy and Stamina
- Maintain Social Participation
- Address Self-Identity Issues

Listen & Understand

Physician Priorities

- Risk of Progression
- PFTs
- HRCT & Extent of Fibrosis
- Identify and Treat Comorbidities
  - GERD
- Develop a Management Plan w/ MDM
- Ongoing Monitoring
- Maximize Survival

Maximize the Patient's Treatment Plan

Overlapping Priorities

- Establish a Strong Patient-Physician Relationship
- Reduce Symptoms
- Choose Effective, Well Tolerated Treatments
- Maximize Quality of Life

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Adapted From: Cheema TJ, et al. Clin Med Insights Circ Respir Pulm Med 2020 March 18;14:1179548420913281.

Patient Centered Communication:

Patient Priorities

- Minimize Uncertainty
  - Understand Disease/Treatments
  - The Scleroderma Foundation: www.scleroderma.org
- Seek Assistance from Partner, Family, & Friends … BUT Maintain Independence
- Maintain Energy and Stamina
- Maintain Social Participation
- Address Self-Identity Issues

Listen & Understand

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- Risk of Progression
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Scleroderma Foundation
scleroderma.org

Summary:

- Screen to identify SSc-ILD
- Monitor closely for progression:
  - Especially those with increased risk and early on in disease
- Treat those:
  - With high risk of progression
  - With clinical symptoms
- Immunosuppressive therapy … consider antifibrotic therapy with ongoing progression
- Develop a multidisciplinary team for the patient
  - PCP and Sub-specialists and Support Teams
- Use patient centered communication to address their concerns with a complex disease and devise an optimal treatment

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Thank You