





## PNEUMOPATHIE INTERSTITIELLE ET **SCLERODERMIE**

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### ATTEINTES PULMONAIRES DE LA SSC

- Très diverses
- PID
   HTAP
   PID + HTP
- PID : cause principale de décès au cours de la SSc
- 1 patient sur 2 ayant une forme diffuse
- 1 patient sur 3 ayant une forme limitée
- → Fréquent et grave : dépistage; diagnostic et traitement de la PID = enjeux centraux

## **DEFINITION DE LA PID-SSc**

- existence de signes radiologiques de PID chez un patient ayant une SSc après avoir exclu les autres causes
- Différent de la FPI : entité radio-pathologique, dans laquelle le rôle de l'histologie pulmonaire est beaucoup plus important.

### PREVALENCE DE LA PID-SSc

- 70-100 % ......données autopsiques
- 30-90 %.....TDM haute résolution **→** 40-50%
- 20-50 % .....radio standard
- 25-43 % ......syndrome restrictif (EFR)

## FACTEURS DE RISQUE D'AVOIR UNE PID-SSC

- Forme cutanée diffuse/Forme cutanée limitée : 2/3 vs 1/3
- Sexe masculin
- Antitopoisomérase 1 : 80% de PID
- Anticentromère : 20% de PID
- Anti RNA POL III : plutôt protecteur (30%)
- Plusieurs associations avec polymorphismes génétiques mais aucun commun avec la FPI (IL1 receptor associated kinase 1, IRF5, MMP12, CTGF, IL-6, CD226, surfactant proteine B, IL-1, HGF et NLRP1)

→ Rien de suffisamment exclusif pour décider de ne pas dépister un patient donné

## PRESENTATION CLINIQUE

- **Asymptomatique** : découverte lors du dépistage +++
- Dyspnée d'effort
- Toux sêche
- Douleur thoracique
- Asthénie
- Râles crépitants +++

### DEPISTAGE DE LA PID

- Systématique au diagnostic de la SSc (PNDS)
- Ne peut dépendre de la présence de signes cliniques ou d'anomalies des EFR/radiographie de thorax/TM6 : trop peu sensibles :
  - 2/3 des patients ayant une PID-SSc ont des EFR normaux initialement
  - 1/2 des patients ayant des EFR normaux ont une PID-SSc
- TDM thoracique en haute résolution +++

CT Features of Lung Disease in Patients with Systemic Sclerosis: Comparison with Idiopathic Pulmonary Fibrosis and Nonspecific Interstitial Pneumonia<sup>1</sup>

• NSIP: 78%

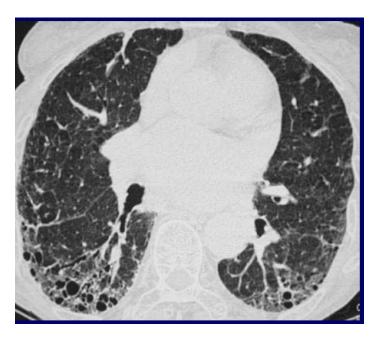
### imagerie:

- ✓ Prédominance de verre dépoli
- ✓ Opacités en bandes
- ✓ Opacités réticulées
  - UIP:8%

#### Réticulations

- √ Cavités en rayon de miel
- √ Bronchectasies par traction
- ñ Réticulations intralobulaires et septale
- ± Hyperdensité en verre dépoli Distribution acinaire périphérique





Desai et al. Radiology 2004

### HISTOIRE NATURELLE DE LA PID-SSc

- Mortalité
- Evolution de la PID-SSc:
  - Scanographique : extension et évolution des patterns
  - EFR

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### **MORTALITE**

#### Causes of death in scleroderma

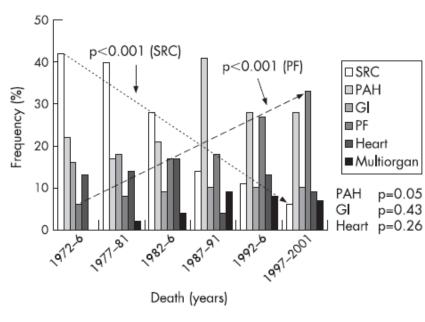


Figure 2 Changes in causes of systemic sclerosis-related deaths between 1972 and 2001. GI, gastrointestinal; PAH, pulmonary arterial hypertension; PF, pulmonary fibrosis; SRC, scleroderma renal crisis.

PID : 1ere cause de décès au cours de la SSc

-33% des décès liés à la SSc

-16% de l'ensemble des décès

### FACTEURS ASSOCIES A LA MORTALITE

- Type histologique?
- TDM haute résolution ?
- EFR ?
- Hypertension pulmonaire ?

### HISTOLOGIE: DISCORDANCE FPI/PID-SSc

80 biopsies pulmonaires : →

- 47 NSIP fibrosantes
- 15 NSIP cellulaires
- •6 **UIP**
- •6 ESL
- •6 divers (bronchiolite, COP)

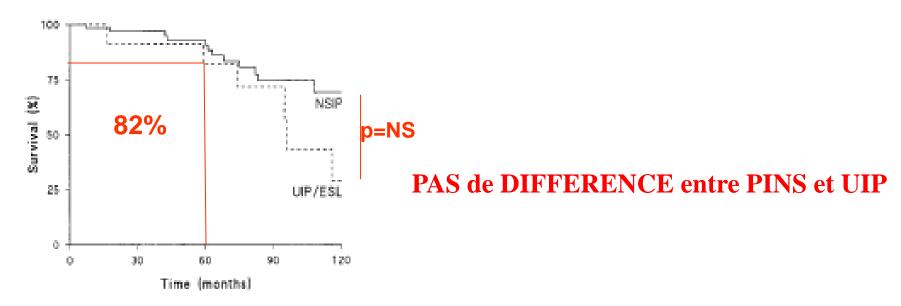


Figure 3. Survival compared between NSIP and UIP/ESL. No significant difference was disclosed.

**PID-SSc** 

### HISTOLOGIE: DISCORDANCE FPI/PID-SSc

### 80 biopsies pulmonaires : →

- 47 NSIP fibrosantes
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- •6 ESL
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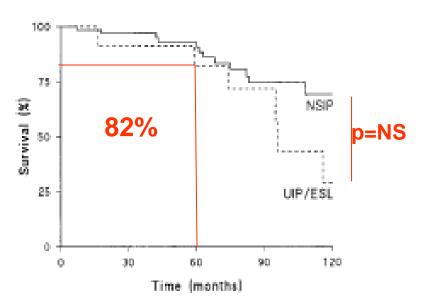
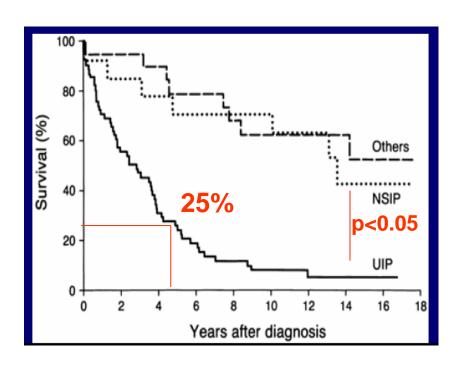


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**FPI** 

**PID-SSc** 

## EXTENSION DE LA PID-SSc

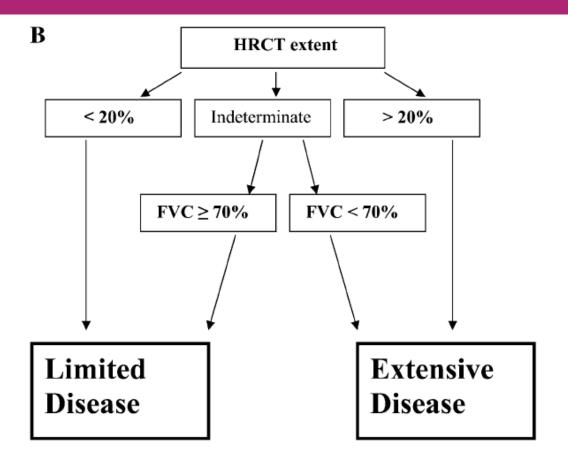
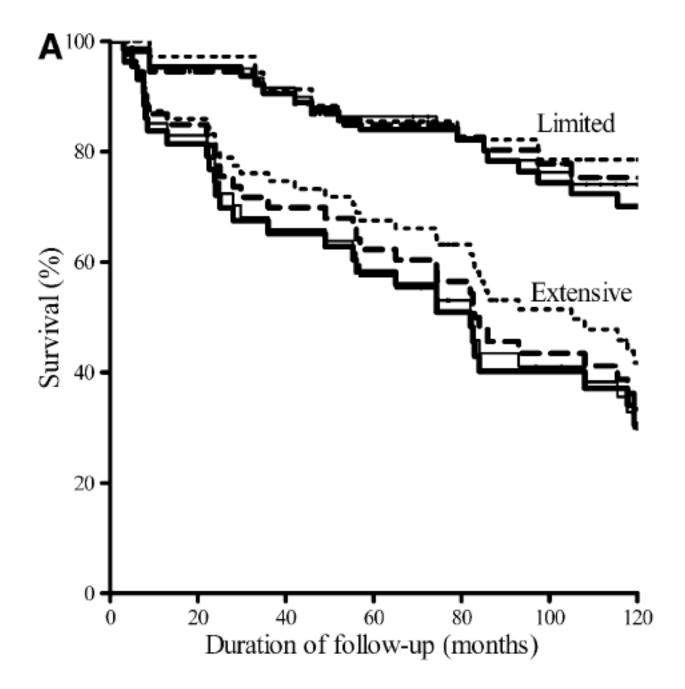


Figure 1. Flow diagram of limited/extensive staging system (A) with the use of formal high-resolution computed tomography (HRCT) scores, for the purposes of analysis, and (B) as applied in clinical practice.



Goh et al. Am J Respir Crit Care Med. 2008

### EFR

• CVF et DLCO : facteurs de risque de mortalité indépendants

Variables	Mortality			
	HR 95% CI p value			
Age	1.03 1.01, 1.05 0.001			
Male Gender	1.98 1.20, 3.26 < 0.01			
Smoking	1.16 0.83, 1.63 0.38			
baseline FEV1	0.98 0.97, 0.99 < 0.01			
baseline FVC	0.99 0.97, 0.99 < 0.01			
baseline DLco	0.95 0.94, 0.97 < 0.0005			
extent of disease	1.03 1.01, 1.04 < 0.0005			
extent of a reticular pattern	1.05 1.03, 1.07 < 0.0005			
proportion of ground-glass	0.99 0.98, 0.99 0.001			
coarseness of reticulation	1.15 1.06, 1.25 0.001			
presence of honeycombing	1.63 1.00, 2.66 0.05			
presence/absence of PHT	4.78 2.68, 8.54 < 0.0005			

## VARIATION DE LA DLCO: FDR indépendant

**Table VII.** Multivariable regression analysis of variables correlated with event (DLCO included in model).

Variable	HR (95% CI)	<i>p</i> -value	
Age at baseline PFT	2.00 (1.99, 2.004)	0.48	
Disease duration at baseline PFT	2.00 (1.99, 2.01)	0.80	
Diffuse disease subtype	1.93 (1.79, 2.07)	0.30	
Anti-Sc170 antibody	1.91 (1.77, 2.06)	0.23	
10% one-year FVC decline	1.94 (1.79, 2.09)	0.43	
15% one-year DLCO decline	1.71 (1.51, 1.91)	0.01	

DLCO: diffusing capacity of the lung for carbon monoxide; FVC: forced vital capacity; ILD: interstitial lung disease; PFT: pulmonary function test.

## ASSOCIATION A L'HTP: FDR indépendant

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Table 5. Multivariate analyses of mortality risk factors in SSc patients who died during the period between the followup HRCT and the end of the study\*

Patient characteristic	HR (95% CI)	P
Pulmonary hypertension, no. (%)	2.0 (1.16-3.52)	0.013
Age at disease onset, years	1.1 (1.06-1.11)	< 0.001
Baseline FVC, %	0.9 (0.97-0.99)	0.017
FVC decline, %	0.9 (0.95-0.99)	0.002
Baseline DLco, %	0.9 (0.97–0.99)	0.019

<sup>\*</sup> Hazard ratios (HRs) and 95% confidence intervals (95% CIs) were estimated by Cox regression analysis using a manual backward elimination procedure. The test of proportional-hazard assumption for the model showed P = 0.590, with all covariates showing P > 0.05. SSc = systemic sclerosis; HRCT = high-resolution computed tomography; FVC = forced vital capacity; DLco = diffusing capacity for carbon monoxide.

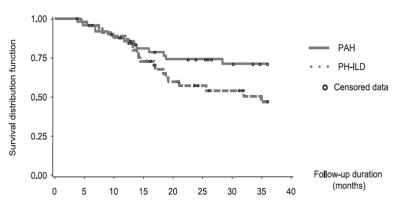


FIGURE 1. Survival in patients with systemic sclerosis (SSc) with PH-ILD or isolated PAH. Survival rates at 36 months were 47% and 71% in the PH-ILD and PAH groups, respectively (log-rank test P=.07). PAH = pulmonary arterial hypertension; PH-ILD = interstitial lung disease-associated pulmonary hypertension.

Author	recruitment period		Survival (95% confidence intervals)
Mukerjee et al. PH-ILD	1998-2002	<b>1</b>	0.50 [ 0.28 , 0.72 ]
Mathai et al. 2009 PH-ILD	2000-2005	-	0.39 [ 0.19 , 0.61 ]
Condliffe et al. PHILD	2001-2006	<b>—</b>	0.28 [ 0.15 , 0.43 ]
Launay et al. 2011 PH-ILD	2001-2006	-	0.47 [ 0.31 , 0.63 ]
Le Pavec et al.	2000-2009	<u></u>	0.21 [ 0.10 , 0.34 ]
PH-ILD 12: 57 % Phet: 0.0532			0.35 [ 0.24 , 0.47 ]

#### PID+HTP: 35% de survie à 3 ans

Hoffmann-Voldt et al. Arthritis Rheum 2015 Lefèvre et al. Arthritis Rheum 2013 Launay et al. Chest 2011

# FACTEURS ASSOCIES A LA MORTALITE RESUME

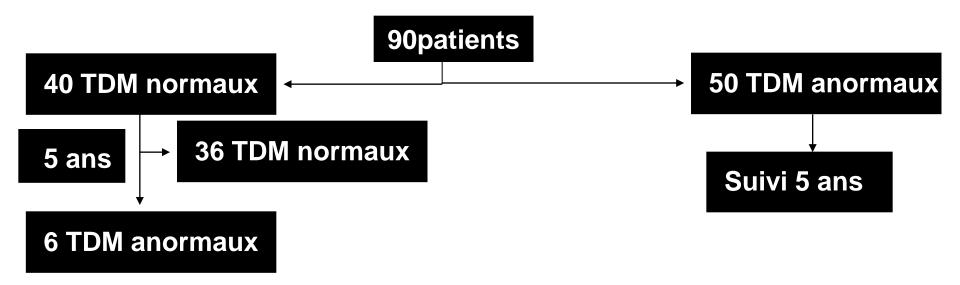
- Type histologique? NON
- TDM haute résolution ? FORME EXTENSIVE
- EFR ? CVF, DLCO, delta DLCO
- Hypertension pulmonaire ? OUI

### HISTOIRE NATURELLE DE LA PID-SSc

- Mortalité
- Evolution de la PID-SSc:
  - Scanographique : extension et évolution des patterns
  - EFR

### EVOLUTION SPATIALE DE LA PID-SSc

- Le plus souvent stable ou lentement progressive
- Progression annuelle : 0,5+/-2,2% du parenchyme pulmonaire
- Variation interindividuelle possible :
  - patients rapidement progressifs
  - Soit d'emblée
  - Soit après une période de stabilité



Crépitants initiaux ACA moins fréquents CV, DLCO initiales plus basses Aggravation de la dyspnée

TDM normale au dépistage : très peu de risque de développer une PID-SSc

### EVOLUTION SPATIALE DE LA PID-SSC MODIFICATION DE PATTERN

- Progression
  - des lobes inf vers les lobes moyens et supérieurs
  - des zones périphériques vers les zones centrales
  - des zones postérieures vers les zones antérieures
- se fait par du VD et des réticulations/bronchectasies pour le poumon sain
- Remplacement du VD existant par réticulations/bronchectasies puis RM
- Pas ou peu de régression du VD : fibrose débutante

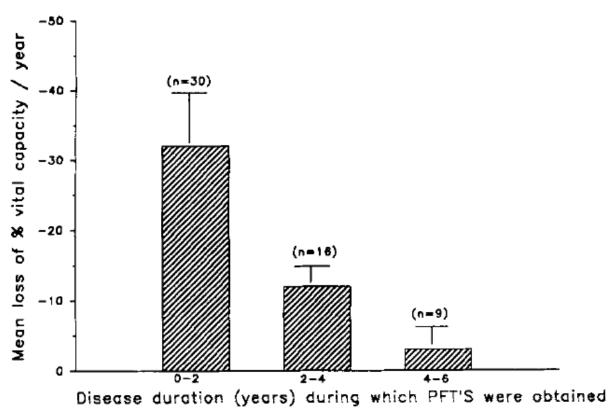


Figure 1. Mean loss of percent vital capacity occurring over 2-year time periods in 55 patients whose initial pulmonary function tests (PFT's) were performed during the first 5 years of scleroderma symptoms.

TABLE 4. SERIAL CHANGES IN PERCENT PREDICTED FVC AND  $DL_{CO}$  LEVELS IN PATIENTS WITH NONSPECIFIC INTERSTITIAL PNEUMONIA, ONE YEAR AFTER PRESENTATION (n = 61) AND 3 YEARS AFTER PRESENTATION (n = 57)

	FVC (1 yr) $(n = 61)$	$DL_{CO} (1 \ yr)$ $(n = 61)$	FVC (3 yr) $(n = 57)$	$DL_{CO} (3 yr)$ $(n = 57)$
Overall change (median) Overall change (range)	-1.7%	-0.3%	-2.5%	-1.8%
	-25.2 to 30.7%	-43.6 to 52.7%	-29.6 to 64.2%	-44.2 to 43.4%
Significant improvement	n = 10	n = 9	n = 13	n = 12
Significant decline	n = 15	n = 8	n = 11	n = 8
Change not significant	n = 37	n = 44	n = 33	n = 37

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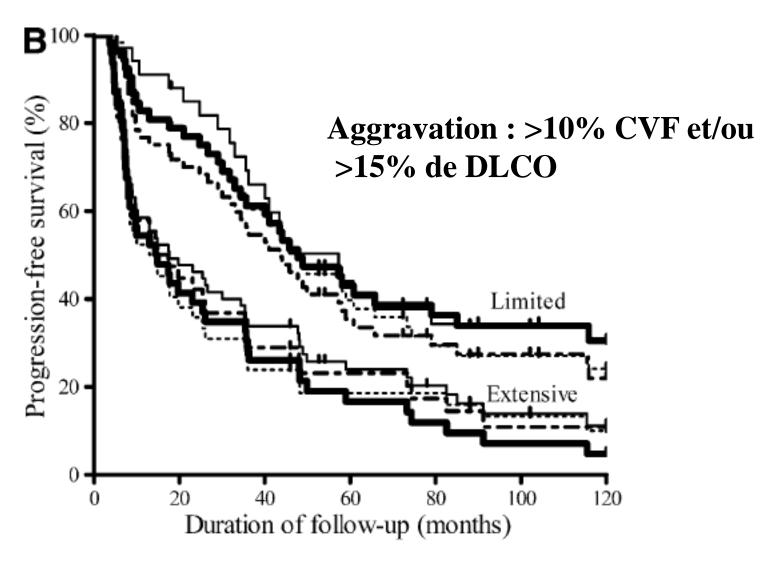
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- Amélioration spontanée des EFR : non observée dans la FPI
- « poussée de fibrose » : 5-10% des patients FPI/an mais moins bien connu dans la SSc-PID

### FACTEURS PREDICITIFS DE L'EVOLUTION DES EFR



Goh et al. Am J Respir Crit Care Med. 2008

### TRAITEMENT DE LA PID-SSC

- Mesures générales
- Traitements immunosuppresseurs
- Autogreffe de cellules souches
- Biothérapie
- Transplantation
- Réhabilitation
- Traitements antifibrosants

### TRAITEMENTS GENERAUX

- Contrôle du RGO +++
- vacciner les patients contre la grippe et le pneumocoque
- Prophylaxie PCP si nécessaire
- Oxygénothérapie



### **CYCLOPHOSPHAMIDE**

SLS-1 FAST

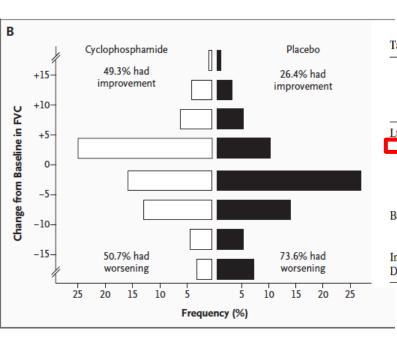


Table 3. Efficacy end point variables*							
	Baseline		1-year followup				
	Treatment group (n = 22)	Placebo group (n = 23)	Treatment group (n = 19)	Placebo group (n = 18)	P†		
Lung function, % predicted							
FVC	80.1 + 10.3	81.0 + 18.8	82.5 + 11.3	78.0 + 21.6	0.08		
DLco <sub>c</sub>	52.9 ± 11.5	$55.0 \pm 12.9$	49.6 ± 10.7	51.8 ± 14.9	0.64		
TLC	$81.8 \pm 10.1$	$76.8 \pm 16.9$	$80.2 \pm 9.8$	$74.4 \pm 16.7$	0.61		
FEV <sub>1</sub>	$79.6 \pm 11.5$	$79.7 \pm 19.1$	$81.3 \pm 12.5$	$77.0 \pm 21.3$	0.16		
Kco	$71.3 \pm 13.4$	$82.7 \pm 19.1$	$71.5 \pm 13.9$	$77.9 \pm 23.3$	0.32		
Baseline HRCT‡							
Disease extent, mean (range) %	20 (6-40)	19 (5-40)	_	_	_		
Ground-glass attenuation, mean (range) %	50 (15–91)	47 (0–95)	_	_	_		
Improvement on serial HRCT, no (%);			6 (40)	3 (20)	0.39		
Dyspnea score, mean (range)§	7.7 (2–14)	7.2 (0–18)	8.75 (0–14)	7.80 (2–14)	0.23		

CVF: + 2.53% (IC: 0.28-4.79) à 12 mois CYC vs placebo

Amélioration modeste de la CVF

Amélioration de la dyspnée et de la qualité de vie dans SLS

Patients essentiellement stables (85% n'ont pas eu besoin de traitement IS)

Tashkin et al. N Engl J Med. 2006 Hoyles et al. Arthritis Rheum. 2006

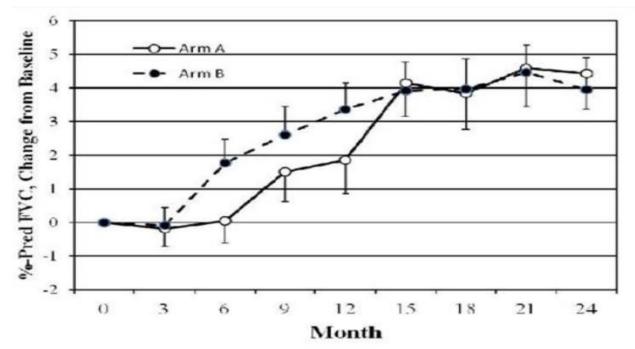
### **MMF en ATTAQUE**

## Etude SLS2

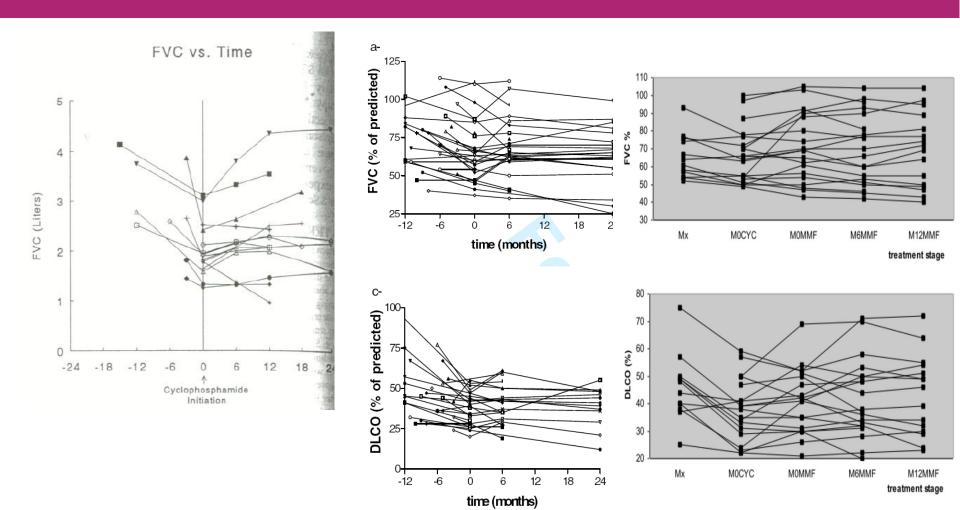
- Etude randomisé en double aveugle
- inclusion:
  - PID-SSc
  - durée d'évolution de la SSc de moins de 7 ans
  - une CVF entre 45 et 80%
  - la présence de verre dépoli sur le scanner thoracique
- randomisation entre CYC 2 mg/kg/j per os pendant 1 an et MMF 1500 mg 2 fois par jour pendant 2 ans.
- Le critère principale de jugement : CVF à 24 mois

#### SLS2

- 142 patients ont été inclus et 106 ont complétés leur visite à 2 ans.
- L'augmentation de la CVF à 24 mois était de 2,88% dans le bras CYC et de 2,15% dans le bras MMF (p=NS entre les 2 mais p<0.05 par rapport à la baseline).



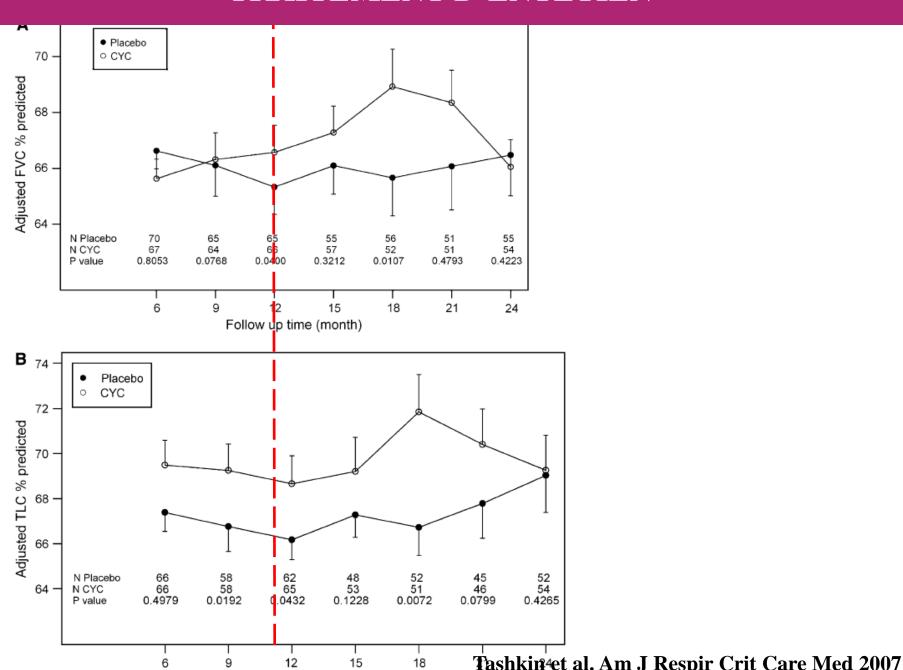
### **CONCEPT: TRAITER LES FORMES AGGRAVATIVES PAR CYC**



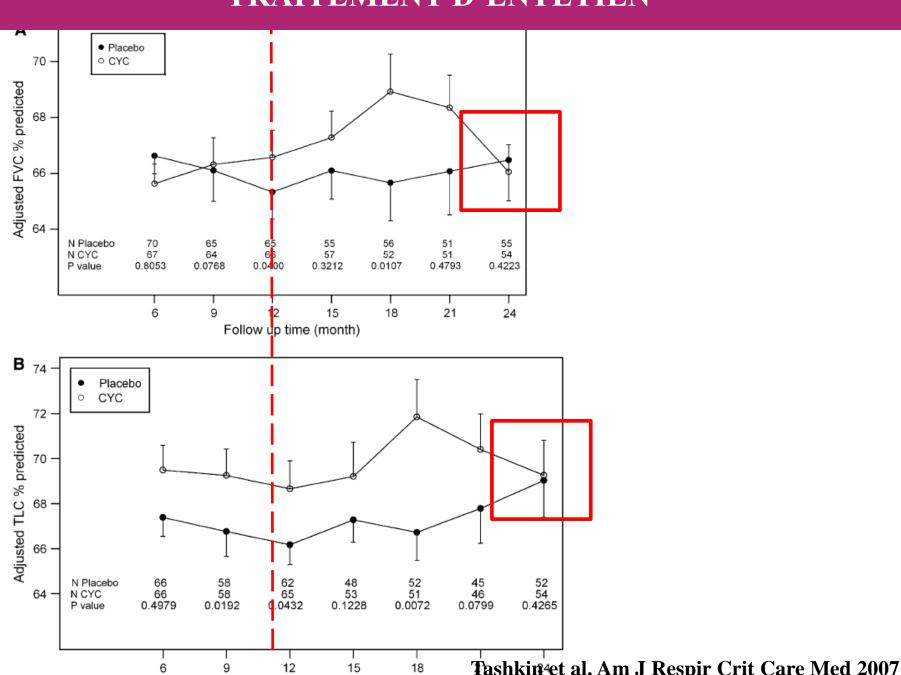
STABILISATION = SUCCES → PHRC SCLEROCYC

Berezne et al. J Rheumatol 2003 Launay et al. J Scleroderma Rel Dis 2016

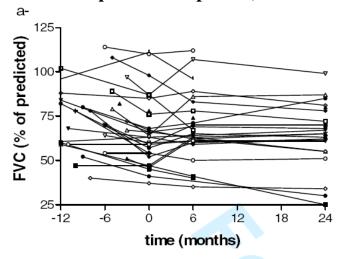
### TRAITEMENT D'ENTETIEN

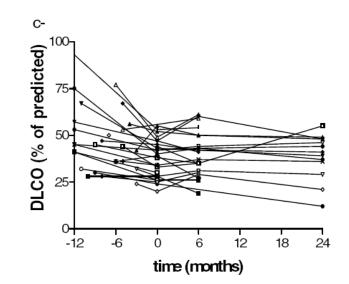


### TRAITEMENT D'ENTETIEN



47% de répondeur après 1,5 ans d'AZA

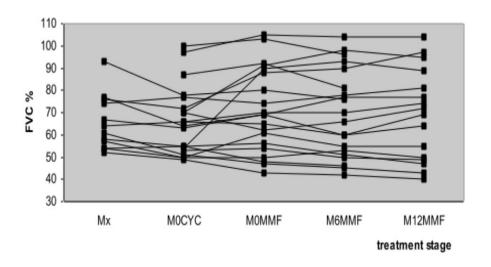


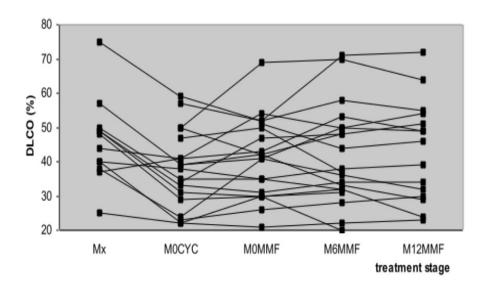


### **AZATHIOPRINE**

Berezne et al. J Rheumatol 2003

55% de répondeur après 1 an de MMF





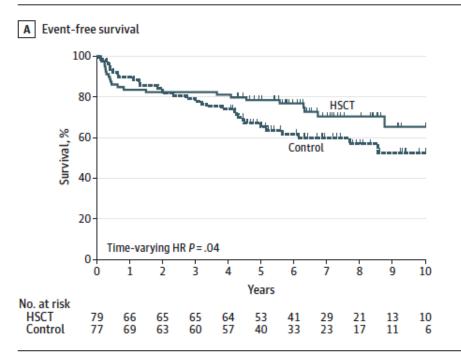
#### **MMF**

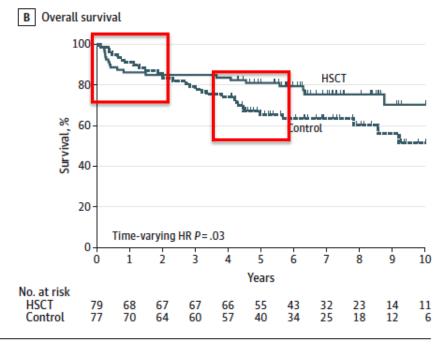
Launay et al. J Scleroderma Rel Dis 2016

# **PERSPECTIVES**

#### **AUTOGREFFE**

Figure 2. Event-Free and Overall Survival During 10-Year Follow-up





Hazard ratios (HRs) and 95% CIs were calculated by Cox regression. Hazard ratios were time-varying. The hazard (slope of the survival curve) in the hematopoietic stem cell transplantation (HSCT) group is initially high because of treatment-related mortality but gradually improves. At 1-year follow-up, the HR already favors the HSCT group, which leads to the crossing of the survival curves at 2 years' follow-up. A, Three-month follow-up: HR, 2.01 (95% CI, 0.74-5.49); P = .17; 6-month follow-up: HR, 1.35 (95% CI, 0.62-2.96); P = .45;

1-year follow-up: HR, 0.52 (95% CI, 0.28-0.96); P = .04; 2-year follow-up: HR, 0.35 (95% CI, 0.16-0.74); P = .006; 3- through 10-year follow-up: HR, 0.34 (95% CI, 0.16-0.74); P = .006. B, Three-month follow-up: HR, 2.40 (95% CI, 0.75-7.67); P = .14; 6-month follow-up: HR, 1.50 (95% CI, 0.61-3.68); P = .38; 1-year follow-up: HR, 0.48 (95% CI, 0.25-0.91; P = .02; 2-year follow-up: HR, 0.29 (95% CI, 0.13-0.65); P = .002; 3- through 10-year follow-up: HR, 0.29 (95% CI, 0.13-0.64); P = .002.

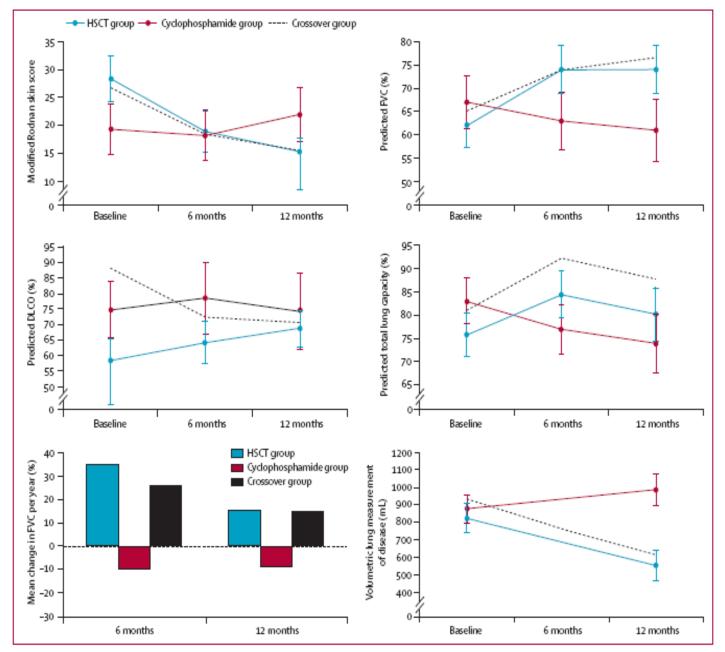
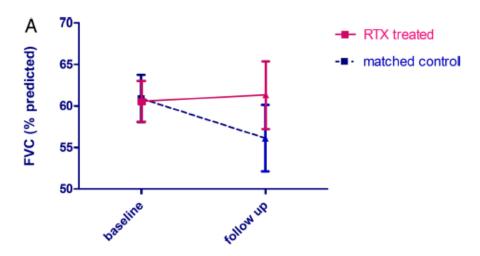


Figure 3: 1-year follow-up for patients receiving haemopoetic stem-cell transplantations, monthly cyclophosphamide, or transplantation after failure of cyclophosphamide

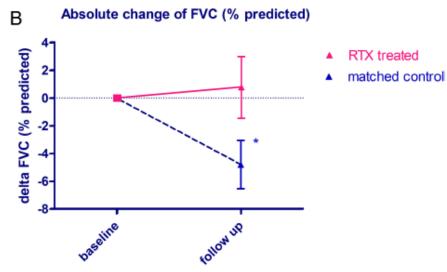
HSCT=haemopoietic stem-cell transplantation. FVC=forced vital capacity. DLCO=diffusing capacity of carbon monoxide.

Burt et al. Lancet 2011

## RITUXIMAB



N=9; RTX (60.6±2.4 B vs. 61.3±4.1 FU;p=0.5) vs. MC (60.9±2.8 B vs. 56.1±4.0 FU;p=0.02)



N=9; 0.8+2.2 vs.-4.8+1.7; p=0.01

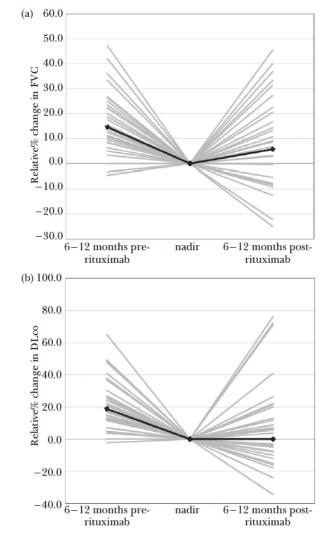
Jordan et al. Ann Rheum Dis 2014

OTHIOLITY IL 7 HITTIGEE

#### Rituximab in severe, treatment-refractory interstitial lung disease

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# **Quarterly Medical Review**



Lung and heart-lung transplantation for systemic sclerosis patients. A monocentric experience of 13 patients, review of the literature and position paper of a multidisciplinary Working Group

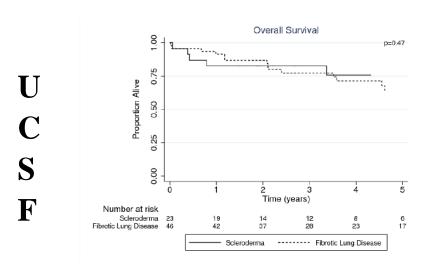
David Launay<sup>1,2,3</sup>, Laurent Savale<sup>4,5,6</sup>, Alice Berezne<sup>7,17</sup>, Jérôme Le Pavec<sup>4,6,8,17</sup>, Eric Hachulla<sup>1,2,3</sup>, Luc Mouthon<sup>7</sup>, Olivier Sitbon<sup>4,5,6</sup>, Benoit Lambert<sup>9</sup>, Marianne Gaudric<sup>10</sup>, Xavier Jais<sup>4,5,6</sup>, Francois Stephan<sup>11</sup>, Pierre-Yves Hatron<sup>1,2</sup>, Nicolas Lamblin<sup>1,12</sup>, Olivier Vignaux<sup>13</sup>, Vincent Cottin<sup>14</sup>, Dominique Farge<sup>15</sup>, Benoît Wallaert<sup>1,16</sup>, Loic Guillevin<sup>7</sup>, Gerald Simonneau<sup>4,5,6</sup>, Olaf Mercier<sup>4,6,8</sup>, Elie Fadel<sup>4,6,8</sup>, Philippe Dartevelle<sup>4,6,8</sup>, Marc Humbert<sup>4,5,6</sup>, Sacha Mussot<sup>4,6,8</sup>, On behalf of the Working Group on Heart/Lung transplantation in systemic sclerosis of the French Network on Pulmonary Hypertension

#### SURVIE DES PATIENTS SCLERODERMIQUES TRANSPLANTES

1.00

#### EXCLUSION DES PATIENTS AVEC I RENALE ET UD ACTIF

#### CRITERES DE SELECTION STRICTS



(2003 – 2007)
R. SAGGAR ET AL.

Column 12 24 36 48 60
Time after lung transplant months

Figure 1. Post-Transplant Survival in Persons with SSc-LD compared to non-CTD-ILD SSc-LD: systemic sclerosis related lung disease, non-CTD-ILD: non-connective tissue disease related interstitial lung disease

**FIGURE 1.** Kaplan–Meier plot displaying the survival estimates for the idiopathic pulmonary fibrosis group (——) versus the systemic sclerosis group (——–). There was no significant difference in overall survival between groups (logrank test, p=0.69).

à 1 an : 83% vs 91% (FPI) à 3 ans : 83% vs 77% (FPI) à 5 ans : 76% vs 64% (FPI)



Sottile et al. Transplantation 2013 Saggar et al. Eur Respir J 2010 Khan et al. Respiratory Med 2013

## TRAITEMENTS ANTIFIBROSANTS

- Nintedanib
- pirfenidone

# Conclusion

- PID-SSc : fréquente et 1ère cause de mortalité
- Rôle majeur du TDM thoracique et des EFR pour le dépistage et l'évaluation de la sévérité
- Importance de l'extension au TDM de la PID-SSc et de l'extension des réticulations
- Stratégie thérapeutique : CYC/MMF dans forme aggravative et forme sévère récente
- Place des antifibrosants dans l'avenir?



IMmune-Mediated INflammatory diseases and Targeted Therapies







