Scientific Abstracts

FRI0371 | CLINICAL ALGORITHMS FOR THE DIAGNOSIS AND PROGNOSIS OF INTERSTITIAL LUNG DISEASE IN SYSTEMIC SCLEROSIS

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Background: Interstitial lung disease (ILD) is currently the primary cause of death in systemic sclerosis (SSc). Thoracic high-resolution computed tomography (HRCT) is considered the gold standard for diagnosis. Recent studies have proposed several clinical algorithms to predict the diagnosis and prognosis of SSc-ILD.

Objectives: To test the clinical algorithms to predict the presence and prognosis of SSc-ILD, and to evaluate the association of extent of ILD with mortality in a cohort of SSc patients.

Methods: Prospective cohort study, including 177 SSc patients assessed by clinical evaluation, laboratory tests, pulmonary function tests, and HRCT. Three clinical algorithms, combining lung auscultation, chest radiography and % predicted forced vital capacity (FVC)[1], were applied for the diagnosis of different extents of ILD on HRCT. Univariate and multivariate Cox proportional models were used to analyze the association of algorithms [1,2] and the extent of ILD on HRCT with the risk of death using hazard ratios (HR).

Results: The prevalence of ILD on HRCT was 57.1% and 79 patients died (44.6%) in a median follow-up of 11.1 years. For identification of ILD with extent $\geq$10 and $\geq$20% on HRCT, all algorithms presented a high sensitivity (>89%) and a very low negative likelihood ratio (<0.16). For prognosis, survival was decreased for all algorithms, especially the algorithm C (HR 3.47, 95% CI 1.62–7.42), which identified the presence of ILD based on crackles on lung auscultation, findings on chest X-ray or FVC <80%. Extensive disease as proposed by Goh and Wells (extent of ILD >20% on HRCT or, in indeterminate cases, FVC <70%) had a significantly higher risk of death (HR 3.42, 95% CI 2.12 to 5.52). Survival was not different between patients with extent of 10 or 20% of ILD on HRCT, and analysis of 10-year mortality suggested that a threshold of 10% may also have a good predictive value for mortality. However, there is no clear cutoff above which mortality is sharply increased.
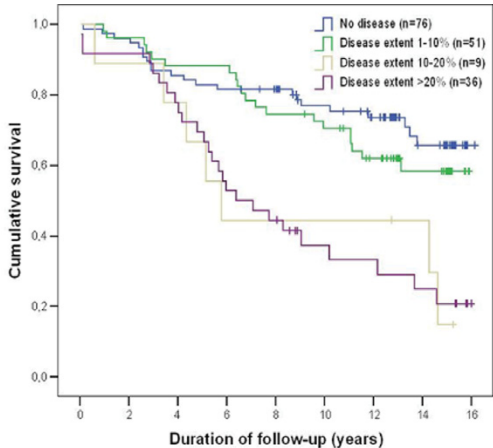

Figure 1. Survival in relation to the extent of lung disease on high-resolution computed tomography (p<0.0001 by log rank test).

Conclusions: Clinical algorithms had a good diagnostic performance for extents of SSc-ILD on HRCT with clinical and prognostic relevance ($\geq$10 and $\geq$20%), and were also strongly related to mortality. Therefore, they probably could be used to obviate the requirement of HRCT in some cases.

References:

[1] Steele R, Hudson M, Lo E, Baron M. Clinical decision rule to predict the presence of interstitial lung disease in systemic sclerosis. Arthritis Care Res (Hoboken) 2012;64:519–24.

[2] Goh NSL, Desai SR, Veeraraghavan S, Hansell DM, Copley SJ, Maher TM, et al. Interstitial lung disease in systemic sclerosis: A simple staging system. Am J Respir Crit Care Med 2008;177:1248–54.

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FRI0372 | THE DUCAS: PROPOSAL FOR A DIGITAL ULCER ASSESSMENT SCORE IN SCLERODERMA

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Background: No objective measure is presently available to assess digital ulcer (DU) in SSc patients apart from "healed/non healed" and experience-based clinical judgment.

Objectives: The aim of the current study is to propose a composite DU clinical assessment score (DUCAS) and, to lend it, test its face validity by correlating it with commonly used disease related patient-reported outcomes (PROs) and physician evaluation.

Methods: SSc patients presenting at least one DU and attending the Rheumatology Wound Care Clinic of the Florence University Hospital or the London Royal Free Hospital were enrolled. Patients were assessed with HAQ-DI, Cochin scale, Visual analogic scale (VAS) for DU-related pain (DU_pain, 0–100 mm), patient VAS for global DU status (ptGDU, 0–100mm) and patient global assessment (PtGA, 0–100 mm) as PROs and physician VAS for DU status (phyGDU, 0–100mm). The DUCAS included 7 DU related variables selected by a committee of 8 SSc DU experts - they are outlined in figure 1. Each variable was weighted on a clinical basis and the DUCAS score was the sum of the values for the 7 variables (max=19.5). Spearman's correlation tests were calculated for to examine face validity. A linear regression model with forward and backward stepwise analysis was used to determine the relationship of individual variables with the primary clinical parameter, phyGDU.

Results: 44 SSc patients (9 males, mean age 54,3±15,6 years, mean disease duration 9,9±5,8 years) were enrolled in the study. Mean phyGDU was 44,3±23mm, mean ptGDU was 54±30mm (Wilcoxon p=0.022, phyGDU VAS vs ptGDU) and mean DUCAS score was 4,2±2. Overall DUCAS showed significant positive correlations with all PROs, but when all the individual clinician and patient's variables were modelled, only the overall DUCAS significantly predicted PhyGDU; after backwards stepwise analysis overall DUCAS and ptGDU best predicted PhyGDU, with an adjusted R^2=0,437 and AIC=380,3 (Table 2).

Table 1

A	Linear Regression for DUCAS		B	Linear Model to PhyGDU			Linear Model to PhyGDU after backwards stepwise		
	Spearman Correlation	p		Estimate	SE	p	Estimate	SE	p
PtGA	0,56	<0,001	PtGA	0,011	0,199	0,955			
PtGDU	0,54	<0,001	PtGDU	0,171	0,233	0,467	0,272	0,101	0,01
DU_Pain	0,44	0,003	DU_Pain	0,048	0,182	0,793			
HAQ-DI	0,44	0,003	HAQ-DI	4,58	7,563	0,549			
COCHIN	0,51	<0,001	COCHIN	0,035	0,252	0,891			
PhyGDU	0,63	<0,001	DUCAS	4,636	1,617	0,007	4,841	1,489	0,002

DUCAS					
1	**Number of Digital Ulcers**			None	0
				1 DU	1
				2 DUs	2
				More than 3 Dus	3
2	**New Digital Ulcers**			YES	1
				NO	0
3	**Gangrene**			YES	3
				NO	0
4	**Surgical approch to DU (above standard care)**			YES	3
				NO	0
5	**Infection of DU**			None	0
				Requiring systemic antibiotics	1
				Osteomyelitis	2
				Septicemia	3
6	**Unscheduled hospitalisation for DU**			YES	3
				NO	0
7	**Analgesics to control DU pain**			No pain	0
				Non requiried analgesics	0,5
				Non-opioids analgesics	1
				Minor opioids	2
				Major opioids	3
				dose increased since last visit	+0,5
				dose decreased since last visit	-0,5
	TOTAL SCORE				

Conclusions: DUCAS is a newly proposed clinical score for SSc related DU which has face validity and which may reflect DU status as judged by SSc experts. Further validation of this score will be undertaken.

Disclosure of Interest: None declared