Researchers develop a risk score to help detect interstitial lung disease in systemic sclerosis patients

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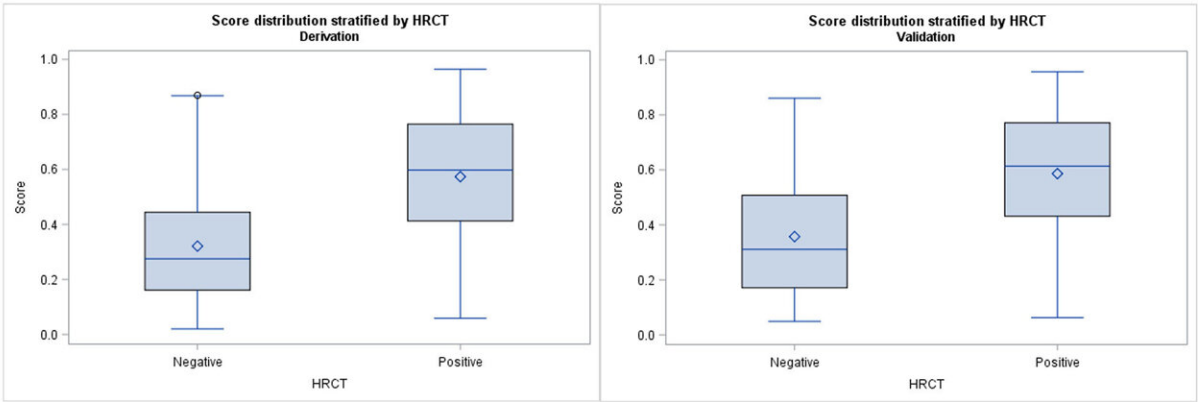
Table 1. Variables included in the final model (A) and performance in the derivation, validation and longitudinal cohorts (B)

A) ILD-RISC MODEL VARIABLES	OR	95% CI	p value
Digital ulcers, ever	2.058	1.347-3.145	<0.001
Age	1.026	1.010-1.042	0.001
SSc_ATB			
Anti-centromere	0.334	0.198-0.563	<0.001
Anti-topoisomerase I	2.379	1.326-4.267	0.004
Anti-RNA-polymerase III	1.407	0.636-3.113	0.399
Anti-Pm/Scl	2.556	0.916-7.135	0.073
None of the above	Comparator		
FVC%	0.990	0.979-1.002	0.091
DLCO%	0.971	0.960-0.982	<0.001
B) ILD-RISC SCORE PERFORMANCE	**Derivation cohort**	**Validation cohort**	**Longitudinal cohort**
All Patients/ILD patients	533/229	247/119	819/170
AUC %, 95% CI	79.1 (75.3 – 83.0)	76.4 (71.0 – 82.7)	72.6 (68.9 – 76.2)
Sensitivity %, 95% CI	85.6 (80.4 – 89.9)	85.7 (78.1 – 91.5)	80.4 (73.9 – 86.0)
Specificity %, 95% CI	53.6 (47.8 – 59.3)	49.2 (40.3 – 58.2)	50.5 (48.2 – 52.9)
Negative Predictive Value %, 95% CI	83.2 (77.2 – 88.1)	78.8 (68.2 – 87.1)	96.3 (94.9 – 97.4)
Positive Predictive Value %, 95% CI	58.2 (52.7 – 63.5)	61.1 (53.2 – 68.5)	13.9 (11.8 – 16.1)

Figure 1. Distribution of the ILD-RISC score between patients with and without ILD on HRCT, in the derivation and validation cohorts.



Credit: Developing a Screening Tool for the Detection of Interstitial Lung Disease in Systemic Sclerosis: The ILD-RISC Risk Score (2022) https://acrabstracts.org/abstract/developing-a-screening-tool-for-the-detection-of-interstitial-lung-disease-in-systemic-sclerosis-the-ild-risc-risk-score/

New research presented this week at ACR Convergence 2022, the American College of Rheumatology's annual meeting, described a first-of-its-kind validated tool to screen for systemic sclerosis-associated interstitial lung disease.

Interstitial lung disease (ILD) is a common complication and cause of death in systemic sclerosis (SSc, scleroderma). Although high-resolution computed tomography (HRCT) is the gold standard for diagnosing systemic sclerosis-associated interstitial lung disease (SSc-ILD), not all patients undergo HRCT screening when first diagnosed with systemic sclerosis.

It is also unclear when the test should be repeated during follow-up of patients who are negative at baseline. The aim of this research was to develop a risk score for the presence of SSc-ILD (the ILD-RISC score) to guide physicians in ordering baseline and follow-up HRCTs.

"Our previous survey identified that only about 66% of physicians regularly screen for interstitial lung disease with HRCT at the time of systemic sclerosis diagnosis, and this percentage drastically drops to less than 15% during follow-up visits," explains Cosimo Bruni, MD-Ph.D., a post-doctoral researcher at the University of Zurich, Research Collaborator at the University of Florence and the study's lead author.

"There are also a certain number of physicians who prefer to be guided by signs, symptoms or other tests to prescribe a chest HRCT. We

therefore wanted to create a tool that guides clinicians about HRCTs, both at diagnosis and follow-up."

The steering board that oversaw the research included six SSc experts, two research fellows and Ilaria Galetti, a patient research partner.

They selected 13 variables considered important in the identification of SSc-ILD: sex, age, time from first non-Raynaud's phenomenon sign or symptom, skin subset, presence of esophageal symptoms, current or past digital ulcers, arthritis, smoking, increased inflammatory markers, New York Heart Association functional class, positive systemic sclerosis autoantibodies, the percent of forced vital capacity (FVC) and diffusing capacity of the lungs for carbon monoxide (DLCO, transfer factor).

The prediction model for ILD was based on baseline visits from systemic sclerosis patients in six European referral centers using multivariable logistic regression with backward selection. Of the 780 included patients, 533 (43% ILD) and 247 (48% ILD) were randomly assigned to the derivation and validation cohorts, respectively.

The researchers combined selected laboratory, clinical and pulmonary function features in their statistical model to obtain the ILD-RISC score. They chose a cut-off with 85.6% sensitivity and 53.6% specificity, which was replicated in the validation cohort and applied longitudinally in a cohort of systemic sclerosis patients with negative baseline HRCT.

"It is important to diagnose ILD early in order to start treatment," Dr. Bruni says.

"We still recommend performing HRCT screening in all patients if the test is readily available and patients are willing to undergo it, so as not to miss any patient with ILD. However, in situations where HRCT is not available, or there are patient concerns about HRCT or the physician

prefers to order it only in selected patients, our ILD-RISC score supports the ability of HRCT to identify this complication while also helping clinicians avoid HRCT when not indicated. Most important, it may help to decide when to order HRCTs at follow-up, thus limiting unnecessary exams."

Dr. Bruni notes the score's relatively low specificity, leading to a certain number of false positives.

"This is a limitation of the current project. We are already working to further improve the score and continue to reduce the number of unnecessary HRCTs," says Dr. Bruni.

More information: Conference abstract

Conference: www.rheumatology.org/Annual-Meeting

Provided by American College of Rheumatology