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Predictors of progression in systemic sclerosis patients with interstitial lung disease

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Lung function tests and chest imaging help predict who has SSc-associated ILD and whether it will progress. In the absence of standardised methods for doctors, we recommend a strategy that combines both lung function tests and chest imaging. http://bit.ly/2uK9ZD2

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ABSTRACT Systemic sclerosis (SSc) is a systemic autoimmune disease affecting multiple organ systems, including the lungs. Interstitial lung disease (ILD) is the leading cause of death in SSc.

There are no valid biomarkers to predict the occurrence of SSc-ILD, although auto-antibodies against anti-topoisomerase I and several inflammatory markers are candidate biomarkers that need further evaluation. Chest auscultation, presence of shortness of breath and pulmonary function testing are important diagnostic tools, but lack sensitivity to detect early ILD. Baseline screening with high-resolution computed tomography (HRCT) is therefore necessary to confirm an SSc-ILD diagnosis. Once diagnosed with SSc-ILD, patients' clinical courses are variable and difficult to predict, although certain patient characteristics and biomarkers are associated with disease progression. It is important to monitor patients with SSc-ILD for signs of disease progression, although there is no consensus about which diagnostic tools

to use or how often monitoring should occur. In this article, we review methods used to define and predict disease progression in SSc-ILD.

There is no valid definition of SSc-ILD disease progression, but we suggest that either a decline in forced vital capacity (FVC) from baseline of ⩾10%, or a decline in FVC of 5–9% in association with a decline in diffusing capacity of the lung for carbon monoxide of ⩾15% represents progression. An increase in the radiographic extent of ILD on HRCT imaging would also signify progression. A time period of 1–2 years is generally used for this definition, but a decline over a longer time period may also reflect clinically relevant disease progression.