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Pleuroparenchymal fibroelastosis in systemic sclerosis: prevalence and prognostic impact

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We provide a thorough assessment of pleuroparenchymal fibroelastosis (PPFE) prevalence, severity and clinical impact in two large cohorts of scleroderma patients (total n=359). PPFE was present in 18% of patients and independently predicted mortality. https://bit.ly/2xDVvG3

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ABSTRACT Interstitial lung disease (ILD) in systemic sclerosis (SSc) is a major cause of morbidity and mortality, mostly presenting as non-specific interstitial pneumonia. Little is known about the prevalence of pleuroparenchymal fibroelastosis (PPFE), a specific entity affecting the visceral pleura and subpleural parenchyma. We set out to estimate PPFE prevalence in two large cohorts of SSc patients and to assess its impact on survival and functional decline.

A total of 359 SSc patients, derived from two referral centres in two different countries (UK and Italy), were included. The first available high-resolution computed tomography scan was independently evaluated by two radiologists blind to clinical information, to quantify ILD extent, freestanding bronchial

abnormalities, and lobar percentage involvement of PPFE on a four-point categorical scale. Discordant scores were adjudicated by a third scorer. PPFE extent was further classified as limited (⩽2/18) or extensive (>2/18). Results were evaluated against functional decline and mortality.

The overall prevalence of PPFE in the combined SSc population was 18% (11% with extensive PPFE), with no substantial difference between the two cohorts. PPFE was significantly linked to free-standing bronchial abnormalities (61% *versus* 25% in PPFE *versus* no PPFE; p<0.0001) and to worse survival, independently of ILD severity or short-term lung function changes (HR 1.89, 95% CI 1.10–3.25; p=0.005).

In the current study, we provide an exhaustive description of PPFE prevalence and clinical impact in the largest cohort of SSc subjects published so far. PPFE presence should be carefully considered, due to its significant prognostic implications.