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ERS/EULAR clinical practice guidelines for connective tissue disease-associated interstitial lung disease

Developed by the task force for connective tissue disease-associated interstitial lung disease of the European Respiratory Society (ERS) and the European Alliance of Associations for Rheumatology (EULAR)

Endorsed by the European Reference Network on rare respiratory diseases (ERN-LUNG)

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This ERS/EULAR clinical practice guideline offers evidence-based recommendations on the screening, diagnosis, monitoring and treatment of CTD-ILDs. It emphasises the importance of further research in areas where evidence is lacking or certainty is low. https://bit.ly/4mX1bPB

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Abstract

Background Interstitial lung disease (ILD) is a frequent manifestation of connective tissue diseases (CTDs) and is associated with high morbidity and mortality. Clinical practice guidelines to standardise screening, diagnosis, treatment and follow-up for CTD-ILD are of high importance for optimised patient care.

Methods A European Respiratory Society and European Alliance of Associations for Rheumatology task force committee, composed of pulmonologists, rheumatologists, pathologists, radiologists, methodologists and patient representatives, developed recommendations based on PICO (Patients, Intervention, Comparison, Outcomes) questions with grading of the evidence according to the GRADE (Grading of Recommendations, Assessment, Development and Evaluations) methodology and complementary narrative questions agreed on by both societies. For both PICO and narrative questions, the Evidence to Decision framework was used to formulate the recommendations.

Results The task force committee concluded with recommendations for 25 PICO and 28 narrative questions, regarding ILD in the context of systemic sclerosis, rheumatoid arthritis (RA), idiopathic inflammatory myopathies, Sjögren disease (SjD), systemic lupus erythematosus (SLE) and mixed connective tissue disease (MCTD). In four narrative questions, regarding screening and assessment of risk for ILD progression in MCTD, SjD and SLE and one PICO question regarding pirfenidone in CTD-ILD other than RA-ILD, the task force had insufficient evidence to support recommendations. Screening, diagnostic, monitoring and treatment algorithms were developed based on the recommendations and usual clinical practice.

Conclusions We provide practical guidance by evidence-based recommendations to clinicians for each of the CTDs. In many cases there is low certainty or absence of evidence and we encourage further research to fill these gaps.