A contemporary practical approach to the multidisciplinary management of unclassifiable interstitial lung disease

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This review describes how patients with unclassifiable ILD should be evaluated and what impact specific clinical, radiological and histopathological features may have on management decisions, focusing on patients with a predominantly fibrotic phenotype https://bit.ly/3o43nqr

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Abstract

Fibrotic interstitial lung diseases (ILDs) frequently have nonspecific and overlapping clinical and radiological features, resulting in ∼10–20% of patients with ILD lacking a clear diagnosis and thus being labelled with unclassifiable ILD. The objective of this review is to describe how patients with unclassifiable ILD should be evaluated, and what impact specific clinical, radiological and histopathological features may have on management decisions, focusing on patients with a predominantly fibrotic phenotype. We highlight recent data that have suggested an increasing role for antifibrotic medications in a variety of fibrotic ILDs, but justify the ongoing importance of making an accurate ILD diagnosis given the benefit of immunomodulatory therapies in many patient populations. We provide a practical approach to support management decisions that can be used by clinicians and tested by clinical researchers, and further identify the need for additional research to support a rational and standardised approach to the management of patients with unclassifiable ILD.