DIFFUSE PARENCHYMAL LUNG DISEASE: AN UPDATE, DIAGNOSIS & MANAGEMENT

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Diffuse Parenchymal lung disease (DPLD) is an umbrella term for over 200 different diseases that display considerable variation in terms of clinical course, treatment and prognosis. Broadly, they can be subdivided into those with an identifiable cause and those without; the latter being referred to as idiopathic interstitial pneumonias. Clinical assessment aims to identify a possible cause; screening for features of systemic disease (eg connective tissue disease) or environmental triggers. Relevant exposures include pneumotoxic drugs, radiation therapy, occupational exposures (eg asbestosis) or implicated allergens (hypersensitivity pneumonitis). Distinguishing the various forms of pulmonary fibrosis is critical for determining correct management and for predicting prognosis. All DPLD is characterized by variable degrees of inflammation and fibrosis. In inflammation dominant disease, the histology is that of organizing pneumonia or non-specific interstitial pneumonia, while in fibrosis dominant disease, Usual interstitial pneumonia (UIP) is present-characterized by fibroblastic foci and only mild to moderate inflammation. These histological patterns are associated with specific radiological features, the recognition of which may abrogate the need for a formal biopsy and tissue diagnosis. With the discovery and approval of two new anti-fibrotic drugs (pirfenidone and nintedanib) heralding a new era in the disease. While these novel antifibrotic agents have been shown to slow the decline in forced vital capacity (FVC), they neither halt progression nor reverse existing fibrosis. These drugs only can be used in fibrosis predominant disease. In inflammation predominant cases can be treated by addressing the etiology. Pulmonary rehab is an important component of management.

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