SSc-ILD: Monitoring and Management Strategies

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Scleroderma-associated interstitial lung disease (SSc-ILD) is a leading cause of morbidity and early mortality, however, its manifestations can range from subclinical chest imaging abnormalities to profound dyspnea and rapidly progressive fibrosis. Identification relies on a combination of clinical risk profiling, chest imaging and pulmonary physiology.

Clinical risk stratification integrates demographic and disease features: early diffuse skin disease, Scl-70 antibody positivity, male sex, and shorter disease duration correlate with a higher risk and faster ILD progression. Screening algorithms increasingly recommend baseline high-resolution computed tomography (HRCT) and pulmonary function testing for all newly diagnosed SSc patients, with more frequent follow-up for those in high-risk subsets. Emerging serum-based biomarkers (serum KL-6, CCL18) are not yet standardized.

HRCT remains a standard for detection and pattern characterization. Pulmonary physiology, particularly forced vital capacity (FVC) and diffusing capacity for carbon monoxide (DLCO), provides a measure of baseline disease severity and allows for longitudinal monitoring. Longitudinal review of respiratory symptoms, pulmonary physiology and 6-minute walk distance (6MWD) provides a practical strategy of monitoring.

Treatment goals are to minimize symptoms, preserve lung function, and prolong survival while minimizing therapy-related drug toxicity. Immunosuppression remains central: cyclophosphamide has shown modest benefit in randomized trials and is commonly used for severe or rapidly progressive disease; mycophenolate mofetil (MMF) is now a preferred first-line agent based on comparative effectiveness and tolerability, showing stabilization or modest FVC improvement over 2 years. Rituximab has reported benefit in observational studies and small trials, particularly for refractory cases. Antifibrotic therapy with nintedanib slows the rate of FVC decline and may be used alone or in combination with MMF. The role of pirfenidone remains investigational. Bone marrow transplantation has also shown benefit in selected patients with rapidly progressive SSc. Lung transplantation is an option for advanced, refractory disease in selected candidates

Holistic care - supplemental oxygen for hypoxemia, pulmonary rehabilitation, vaccination to prevent respiratory infection, management of gastroesophageal reflux, and identification/treatment of pulmonary hypertension - remains crucial. The common presence of adverse treatment effects necessitate routine monitoring.

In summary, SSc-ILD management now combines standardized screening and longitudinal monitoring, immunomodulation and antifibrotic therapy, and individualized holistic care.