Defining Subsets In Systemic Sclerosis-Interstitial Lung Disease

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INTRODUCTION

- Systemic sclerosis associated interstitial lung disease (SSc-ILD) patients have variable severity, risk for progression, and progression of lung disease.
- Cohort enrichment strategies and treatment algorithms depend on shared definitions of these SSc-ILD subsets.
- No consensus-based definitions exist that delineate these SSc-ILD subsets.

OBJECTIVE

- Identify patient and disease-related features most important to ILD experts in the classification of severity, risk for progression, and progressive SSc-ILD subsets.

METHODS

- Eighty profiles were created: Scleroderma Lung Study-II (n=53) & Michigan Cohort (n=27).
- Each profile contained demographics, disease features, and assessments of disease severity.

EXPERTS

- Eighty-eight international experts were contacted to participate in an on-line survey.
- Each expert received 10 patient profiles: 5 baseline and 5 baseline with 1 year follow-up.
- Baseline profiles were rated on severity and risk for progression, follow-up on progression.
- Each profile was rated by at least 4 experts; consensus was 75% concordance.

RESULTS

- Our group successfully created real-world cases of SSc-ILD patients into concise patient profiles.
- A majority consensus in classification of disease severity, risk of progression, and progression was met.
- Patient and disease-related features important to experts were identified for each domain.
- For disease severity, a CART analysis identified features (WLI & DLco) most important in classification.
- These data represent the first step in creating operational definitions for ILD subsets.

CONCLUSION

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