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Title: The impact of definition of ILD on WHO group Classification of PH in scleroderma

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Body: Background:Patients with scleroderma (SSc) can develop several forms of pulmonary hypertension (PH). However, while guidelines for classification of PH distinguish between WHO Group I and II disease, they fail to distinguish between PH-ILD [group III] and PAH with thresholds of pulmonary function or chest radiographic findings that define PH-ILD. Therefore, we applied various definitions of “significant” ILD to our population of SSc patients with known PH to determine the impact on PH classification. Methods: 150 consecutive patients with SSc and either Group I or III PH documented by RHC were retrospectively reviewed. We applied 3 definitions of “significant” ILD: 1) TLC<60% predicted or TLC 60-70% predicted with > “minimal” fibrosis (clinical trial definition) 2) Limited/Extensive staging system: extent of lung involvement>20% of lung fields or <20% with FVC<70% predicted 3) FVC<70% predicted. Patients were then reclassified as Group I or III based upon these criteria. Results: Using the clinical trial definition, 73% of patients had Group I disease; however, when applying the limited/extensive staging system or FVC criteria, only 39% patients had Group I disease. No differences in demographics or hemodynamics were noted between groups based upon ILD definition. Conclusions: We found a substantial impact upon the WHO group classification of patients with SSc-PH based upon criteria used to define ILD with changes in >40% of patients. Given the differences in response to therapy and outcomes in SSc between Group I and Group III disease, the changes in PH classification based upon definition of ILD demonstrated in this study have important implications for clinical trials and clinical practice.