# PULMONARY MANIFESTATIONS IN A COHORT OF EGYPTIAN SYSTEMIC SCLEROSIS PATIENTS

Thesis

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Bu

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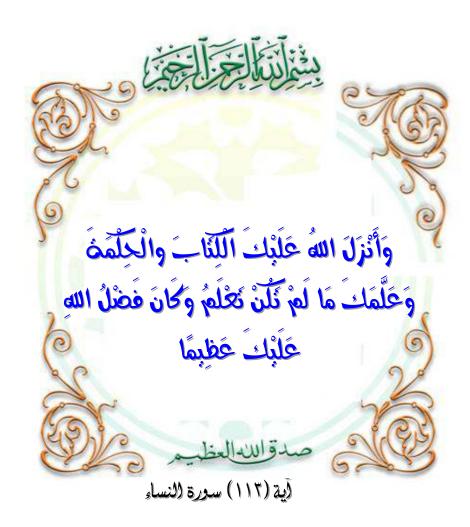
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### List of Abbreviations

#### Full term Abb. 6MWT ...... Six Minute Walk Test ACE..... Angiotensin Connerting Enzyme ACR...... Americal Collegue of Rheumotolology AZA ..... Azathioprine CCBs ...... Calcium Channel Blockers CD ...... Cluster of Differentiation COMP...... Cartiage Oligomeric Matrix Protein COPD ...... Chronic Obstructive Pulmonary Disease CRP...... C-Reactive Protein CTGF...... Connective Tissue Growth Factor CXCL4..... Chemokine (C-X-C motif) Ligard 4 CYC ...... Cyclophophaamicle dcSSc...... Diffuse Cutaneous Systemic Sclerosis DLCO ...... Diffusion Lung Capacity for Carbon Monoxide ECG..... Electrocardiogram ECM ..... Extracellular Matrix ESR ..... Erythrocyte Sedimention Rate ESRD ..... End Stage Renal Disease EULAR..... European League against Rheumatism EUSTAR ..... Europeay Sclearodrma Trials and Research FEV1..... Forced Expiratory Volume in First Second FVC ...... Forced Vital Capacity

### List of Abbreviations cont...

#### Full term Abb. GERD......Gastroesophageal Reflux GGOs..... Ground Glass Opacities GI ..... Gastrointestinal HRCT ...... High Resolution Computed Tomography HSCT ...... Hematopoietic Stem Cell Transplantation ILD..... Interstitial Lung Disease lcSSc..... Limited Cutaneous Systemic Sclerosis LUS ..... Lung Ultrasound MI...... Myocardial Infarction MIP-1a ...... Macrohpage Inflammatory Protein-1a MMF...... Mycophenolate Mophetil mPAP ...... Mean Pulmonary Artery Pressure MRSS ...... Modified Rodnan Skin Score MTX ...... Methotnexate NSAID...... Non Steroid al Anti-Inflammtory Drugs NSIP...... Non Specific Interstitial Pneumonia NTproBNP...... N-Terminal Pro-Brain Natriuretic Peptide OP ...... Organizing pneumonia PDE..... Phosphodiesterase PDGF ...... Platelet Derived Growth Factor PFTs.....Pulmonary Function Tests PH ...... Pulmonary Hypertension

### List of Abbreviations cont...

#### Full term Abb. PMN.....Polumorphnuclear PVOD ...... Pulmanory Veno-Occlusive Disease RA ..... Rheumatoid Arthritis RCTs ...... Randomised Controlled Trials RHC ..... Right Heart Catherization RP..... Raynaud's Phenomenon RTX ..... Rituximab SRC ..... Scleroderma Renal Crisis SSC..... Systemic Scleorsis TGF-B ..... Transforming Growth Factor Beta TGF-β..... Transfoming Growth Factor-β TLC ...... Total Lung Capacity TNFa..... Tumor Necrosis Factor a UIP..... Usuall Interstitial Pneumonia VEDOSS ...... Very Early Diagnosis of Systemic Sclerosis

#### **ABSTRACT**

Background: Systemic sclerosis (SSc) or Scleroderma is a multisystem autoimmune disease of unknown etiology that is characterized by endothelial dysfunction resulting in vasculopathy of small vessels, dysfunction of fibroblasts with resultant excessive collagen production and fibrosis. Skin affection in SSc is nearly a universal feature. While almost all patients with SSc have issues with their skin, particularly extreme scarring, swollen, tight, or hard skin on their fingers, numerous also have tight, swollen, or hard skin in other body areas, especially the face and the arms. They additionally have systematic affection, fundamentally the vascular and the immunologic system. Pulmonary disease is a crucial component of SSc. It is estimated that about 80% of patients with SSc have some evidence of pulmonary affection. So, pulmonary disease comes as the most frequently seen visceral component second only to esophageal disease. Also, pulmonary affection predicts a poorer prognosis in patients with SSc.

**Objective:** The aim of this work was to study the occurrence of the pulmonary manifestations especially Interstitial Lung Disease (ILD) and Pulmonary Hypertension (PH) in a cohort of Egyptian patients with SSc and correlation with the extent of skin affection.

**Methods:** A cross-sectional, observational study involving 30 adult patients diagnosed with systemic sclerosis fulfilling 2013 ACR/EULAR Classification Criteria for Scleroderma was done.

**Results:** All our studied patients (100%) showed some degree of restriction in the pulmonary function tests. 25 patients (83%) showed evidence of ILD by the HRCT of the chest. 5 patients (17%) had evidence of PH in the echocardiography. There was a significant relation between degree of skin affection and some manifestations of ILD especially the grade of dyspnea. There was a statistically significant relation between degree of skin affection and severity of restriction on Pulmonary Function Tests, presence of PH in Echocardiography. There was also a statistically highly positive correlation between degree of skin affection and the degree of ILD in the HRCT of the chest.

Conclusion: SSc is a progressive multisystem disease. Females are more affected than males. Pulmonary manifestations are very common in SSc. The most common pulmonary manifestations are ILD and PH. Patients with combined ILD and PH had a greater duration of the disease than those with ILD only. Patients with diffuse cutaneous SSc have more manifestations and more severe disease than those with limited cutaneous SSc. There is a strong relation between skin involvement and visceral involvement particularly pulmonary involvement, but this need to be confirmed by further larger studies.

**Key Words:** Systemic Sclerosis (SSc), Interstitial Lung disease (ILD), Pulmonary Hypertension (PH).