

European Respiratory Society Annual Congress 2012

Abstract Number: 2062

Publication Number: P3932

Abstract Group: 4.3. Pulmonary Circulation and Pulmonary Vascular Disease

Keyword 1: Pulmonary hypertension **Keyword 2:** Skeletal muscle **Keyword 3:** Exercise

Title: Iron deficiency in patients with systemic sclerosis-associated pulmonary arterial hypertension

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Body: Background Systemic sclerosis-associated pulmonary arterial hypertension (SSc-PAH) has a poor clinical outcome compared to other types of PAH. Recent data has shown that iron deficiency (ID) is associated with poor survival in idiopathic PAH. Inflammatory cytokines and increased hepcidin levels play a role. We hypothesise that a high prevalence of ID in SSc-PAH is linked to poor clinical outcome. Methods Measures of body iron status were performed retrospectively in serum from SSc-PAH patients (n=49) and systemic sclerosis patients without PAH (SSc, n=131). Six minute walking distance (6MWD) was also compared between the groups. Results Circulating soluble transferrin receptor (sTfR) levels in SSc-PAH patients were higher than in SSc (p<0.001) while circulating iron, ferritin and transferrin saturation were reduced. The prevalence of ID, defined by sTfR >28.1 nmol/L, was 47% in SSc-PAH compared to 20% in SSc (p<0.001). Although hepcidin levels were lower in SSc-PAH than in SSc patients (p<0.001), hepcidin in both groups were high compared to reference values. There was no significant correlation with interleukin-6 (IL-6) levels (p=0.82), since IL-6 was higher in SSc-PAH compared to SSc patients (p<0.01). 6MWD was lower in SSc-PAH compared to SSc patients (p<0.001) and was even more reduced in case of ID (SSc-PAH with ID vs SSc-PAH without ID, p<0.05). Conclusions Iron deficiency is more prevalent in SSc-PAH than in SSc patients and is associated with lower exercise capacity. The role of hepcidin in this process remains to be elucidated.