RESPIRATORY MUSCLE WEAKNESS AS A CAUSE OF EXERCISE INTOLERANCE IN PATIENTS WITH B-THALASSEMIA MAJOR

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Background: Patients with β-Thalassemia major (TM) are characterized by a decreased exercise capacity. Respiratory muscles and their relation with exercise capacity have not been evaluated in these patients yet.

Materials and Methods: Our study population consisted of 8 patients (4M/4F) with TM (mean age: 27.7±7.8 yrs) and 8 apparently healthy age-matched control subjects (8M, 34.4±10.8 yrs). All patients with TM had regular transfusions and chelation with desferoxamine. None had a history of chronic pulmonary disease. All individuals (n=16) performed spirometry and maximal inspiratory pressure (PImax) was also measured. Then the whole population underwent a symptom-limited incremental cardiopulmonary exercise testing. Measurements included peak O_2 uptake (VO_2p), anaerobic threshold (AT) and ventilatory response to exercise (V_E/VCO_2 slope).

Results: Patients with TM had lower % predicted FEV_1(77±12 vs 105±18, p=0.003) and % predicted FVC(82±12 vs 108±21, p<0.01) than controls, but the ratio FEV_1/FVC had no difference between the two groups (81.1±4.9 vs 80.6±6.7, p=ns). TM patients had decreased VO_2p (21.6±5.3 vs 29.9±8.3 ml/kg/min; p<0.05) and lower AT (14.5±3.4 vs 19.8±6.4 ml/kg/min; p=0.05) than controls. No difference was observed at the ventilatory response to exercise as expressed by the V_E/VCO_2 slope, as well as peak HR. PImax was correlated significantly with VO_2p (r:0.8, p<0.001).

Conclusion: Our data indicate that patients with TM have a restrictive pulmonary pattern and reduced exercise capacity. This exercise intolerance seems to be strongly related to respiratory muscles abnormalities which may be explained by systemic adverse effects related to disease and its therapy.