

STRENGTH, MUSCLE QUALITY AND FUNCTIONAL CAPACITY IN LIVER TRANSPLANTED FAMILIAL AMILOIDOTIC POLINEUROPATHY PATIENTS

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Introduction: Liver transplantation is the unique treatment for several end stage diseases. Familial Amiloidotic Polineuropathy (FAP) is a neurodegenerative disease related with systemic deposition of amyloid fibre mainly on peripheral nervous system, clinically translated by an autonomous sensitive-motor neuropathy with severe functional limitations in some cases. The unique treatment for FAP disease is a liver transplant with a very aggressive medication to muscle metabolism and force production. To our knowledge there are no quantitative characterizations of body composition, strength or functional capacity in this population. The purpose of this study was to compare levels of specific strength (isometric strength adjusted by lean mass or muscle quality) and functional capacity (meters in 6 minutes walk test) between FAP patients after a liver transplant (4.1±2 months after transplant surgery) (FAPT) and a healthy group (HG). Sixty-four subjects were assigned in 2 groups: 46 patients FAPT (27 males, 32±8 yrs; BMI 21.8±3.8 and 19 females, 37±5 yrs; BMI 22.4±4.0) and eighteen HG (9 male, 34±7 yrs; BMI 24.3±1.8 and 9 female, 36±8 yrs; BMI 23.2±2.6). Isometric strength of quadriceps was measured using an isokinetic dynamometer (Biodex). Body composition was determined by measuring lean mass of dominant lower extremity in a region of interest (thigh) by dual-energy x-ray absorptiometry. Muscle quality was ascertained by taking the ratio of strength to muscle mass. Results: HG showed significant higher values than FAPT patients for: peak torque (66.3N±25.2N vs 40.2N±17.6N respectively p=.000), muscle quality (11.8±2.6 vs 7.9±2.7 respectively, p=.000) and functional capacity (675.8m±109.1m vs 511.4m±139.1m respectively, p=.000). There are no differences between HG and FAPT for BMI (23.8±2.2kg/m² vs 22.0±3.8 kg/m² respectively, p=.07) and thigh muscle mass (5.5kg±1.2kg vs 5.05kg±1.1kg respectively p=.14). Negative correlations were observed for FAPT patients but not for HG between age and peak torque (p=.013; r=-.363) and age and thigh muscle mass (p=.011; r=-.373). When comparing functional capacity, peak torque, muscle quality and thigh muscle mass between males and females in each group significant higher values were observed (p<.05) in males for HG but these differences disappeared in FAPT patients with exception for thigh muscle mass (p=.000; 5.6kg±1.02kg vs 4.3kg±0.7kg for males and females respectively). Conclusions: FAPT patients have lower functional capacity, strength and muscle quality than HG. The differences between groups for muscle quality and peak torque but not thigh muscle mass seems highlight the importance of the neural component of the disease and show also the importance of training the process of force production specially the sensoriomotor component in FAP patients and probably the importance of a strengthening exercise program. Further studies are needed to explore training effects on function after transplantation.