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Amyloidotic Polyneuropathy (FAP): The relevance of muscle mass

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Loss of muscle mass and function is a common occurrence in liver transplant Familial Amyloidotic Polyneuropathy (FAP) patients. Sarcopenia is associated with morbidity and mortality before and after liver transplantation. However, the ability of skeletal muscle mass to recover after transplant remains uncertain, and so as the the importance of clinical exercise prescription.

METHODS

Participants were 39 FAP patients (22 males) aged 23-59 years, who had been submitted to a liver transplant (Tx) 2-4 months earlier. Sarcopenia was defined according to the criteria of the International Working Group on Sarcopenia and Society of Sarcopenia, Cachexia and Wasting Disorders¹.

Whole-body dual X-ray absorptiometry (DXA) was used to measure body fat and leansoft tissue.

From this data, appendicular skeletal muscle mass (kg) was calculated by summing the muscle mass of the four limbs, assuming that all non-fat and non-bone mass is skeletal muscle. Skeletal Muscle Index (SMI) was calculated adjusting appendicular skeletal muscle mass to the squared height.

Functional aerobic capacity was assessed using the 6 min walk test (6MWT) and handgrip strength was measured on the dominant hand using a hand dynamometer.



RESULTS

Table 1 – Characterization of sample (22 males; 17 females)

VARIABLE	Males (22)	Females (17)
Age (years)	31.7±7.9	37.1±5.0
Time Post-Tx (month)	4.1±1.9	3.4±1.9
Weight (Kg)	67.7±13.2	56.8±10.6
Height (m)	1.7±0.1	1.6±0.1
BMI	22.4±4.0	21.6±3.9
Fat Mass (%)	18.7±7.4	27.8±7.6
Total Skeletal Muscle Mass (TSMM)(kg)	25.5±4.3	17.2±2.4
Skeletal Muscle Index (SMI)	7.2±1.0	5.7±0.8
Right Handgrip	37.4±11.8	27.7±7.9

Fig. 2 – SMI distribution in males

Fig. 3 – SMI distribution in females

The prevalence of sarcopenia according to SMI values was 45,5% in males and 41,2% in females. According Dunn (2017)³ diminished SMI is strongly associated with increased hospitalizations and cost of care In addition, 27,3% of males and 17,6% of the females could be classified as having sarcopenia with low mobility (distance < 400m).

Fig. 3 –6MWT distribution by gender.

6MWT (m)

514.3±121.4 507.7±150.4

Legend: BMI - Body Mass Index; TSMM acording equation of Kim, Wang et al, 2002²; SMI = Appendicular Skeletal Mass adjusted to squared height in meters; 6MWT - 6 minutes walk test

A fat mass higher than 16% for males and 26% for females was found in 54,5% of males and 52,9% of females.

Values for low handgrip strength related with sarcopenia definition were found in 27,3% of males (handgrip<30kg) and in 17,6% of females (handgrip<20kg).

Also for this population a strong association was verified between handgrip strength and SMI.

DISCUSSION and CONCLUSIONS

Sarcopenia seems to have a high prevalence in liver transplanted patients and could be also prevalent in FAP patients due to essentially previous malnutrition problems associated to this disease. Liver transplant seems to increase its prevalence, probably because of the aggressive medication with impact on muscle metabolism. However the association between sarcopenia in FAP patients with or without liver transplant was not studied yet. Future studies should examine the efficacy of nonpharmacological, multimodal approaches, such as exercise training and a nutritional intervention, that may substantially reduce the functional decline in TX FAP.

References

1.Cruz-Jentoft et al. Sarcopenia: European consensos on definition and diagnosis. Age and ageing; 2010; 39: 412-423; 2. Kim, J.; Wang, Z et al. Total-body skeletal muscle mass: estimation by a new dual-energy X-ray absorptiometry method.; Am J Clin Nutr, 2002 Aug;76(2):378-83; Dunn, MA The cost of sarcopenia. Transpl Int. 2017 Sep 14. doi: 10.1111/tri.13069.

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