

**137****BODY FAT ASSESSMENT BY BIOELECTRICAL IMPEDANCE IN PATIENTS WITH MUCOPOLYSACCHARIDOSIS**

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Background: Mucopolysaccharidoses (MPS) are lysosomal disorders characterized by the deficiency in lysosomal enzymes in the degradation of glycosaminoglycans. Abnormal accumulation of this molecule compromises cellular and organic function, leading to a spectrum of clinical manifestations, both multisystem and progressive. Despite not being a disease for which a dietary treatment will modify the outcome, it is a condition in which patients benefit from nutritional monitoring. Objective: To evaluate the nutritional status of patients with MPS seen in a specialized outpatient, by means of body composition assessment by bioelectrical impedance (BIA). Methods: A cross-sectional study with a convenience sampling. Inclusion criteria were confirmed diagnosis of MPS, patient with  $\geq 10$  years of age and the presence of conditions appropriate for the performance for the exams. Subjects were evaluated by means of anthropometrics and body composition parameters provided by BIA. Results: thirteen patients were enrolled, seven (53.8%) females and six (46.2%) males, with a median age of 22 (12-28) and 15 (10-19) years, respectively. Regarding to nutritional status, five (38.4%) were eutrophic, four (30.8%) were overweight, and four (30.8%) were obese. There was no statistical difference in the percentage of fat-free-mass (FFM) and fat mass (FM) when compared to different types of MPS. However, it was noted that patients with MPS IVA presented with a greater tendency in %FM in relation to the other MPS types studied. Enzymatic replacement therapy (ERT) patients showed a %FM statistically lower compared to the group without ERT, suggesting a possible treatment effect on body fat accumulation. Conclusion: We believe it is very relevant in nutritional monitoring management of patients with MPS, aiming to adapt the nutritional parameters. Additional studies with larger samples should be conducted in an attempt to confirm this hypothesis. Palavra-chave: Mucopolysaccharidoses; Bioelectrical Impedance; Enzyme Replacement Therapy. Projeto 12-0259