P-035 - NUTRITIONAL STATUS IN PATIENTS WITH MUCOPOLYSACCHARIDOSIS TYPE I AND II.

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INTRODUCTION: There is scarce information on body composition in Mucopolyssacharidosis (MPS), and how the accumulation of glycosaminoglycans (GAG) affects it. The aim of this study was to investigate the nutritional and metabolic aspects of adult male patients with MPS types I and II assisted at Reference Center for Inborn Errors of Metabolism at Universidade Federal de São Paulo. METHODS: The body composition analysis was made by plethysmography, assessment of dietary intake by food recording 24h (three nonconsecutive days), resting metabolic rate by measuring oxygen consumption and energy requirements estimated by prediction equations. Anthropometric results (weight, height, body mass index) were compared with WHO references. RESULTS: Five patients (3 MPS I / 2 MPS II) who were under enzyme replacement therapy (ERT) were included. The mean age at diagnosis and at the time of the study was 18.6 years (14y – 27y) and 24.1 years (23y – 30y, respectively. The mean time under ERT was 3.5 years (1.1y - 5.8y). The mean value of resting metabolic rate was 1386 kcal/day (967 kcal/day - 1.803 kcal/day): one low, two normal and two high. The estimated mean energy requirement was 2240 kcal/day (1642 kcal/day - 2447 kcal/day). Blood glucose and total cholesterol levels of all patients were within the normal reference range. Serum high-density lipoprotein levels (HDL) were below the recommended in all patients. The evaluation of body composition by plethysmography showed that 3 out of 5 patients had about 9% fat (lean), one about 14.3% (moderately lean) and one 20.5% (overweight). Patients consumed macronutrients within the normal range distribution rate. There was no difference in body mass index between the patients with the mean 22.6 kg/m2(20.4-24.0kg/m2), which does not corroborate with the Brazilian population with more than 48.8% overweight according to the Brazilian Institute of Geography and Statistics (IBGE). CONCLUSION: In this study, patients with MPS did not present significant alterations in the analyzed parameters, however, these results should be interpreted with caution since the influence of GAGs in the result of the body composition method used is not totally understood.