P-017 - EVALUATION BY ANTHROPOMETRY OF THE NUTRITIONAL STATE OF PATIENTS WITH INBORN ERRORS OF METABOLISM IN PROTEIN-CONTROLLED DIETS

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INTRODUCTION: The nutritional treatment of the inborn errors of metabolism (IEM) of amino acids consists of limiting the supply of natural proteins and complementing it with specific formulas that exclude the toxic amino acid/s. It is known that through a protein-controlled diet (PCD), favorable results are obtained in the medical treatment, but there is little information about their impact on growth. OBJECTIVES: To evaluate the nutritional state of patients with IEM that carry out protein restricted diets, through routine anthropometric measurements. To know the impact of said diets on the nutritional state.

MATERIALS AND METHODS: Cross sectional study from patients followed by the pediatric metabolic service at the Hospital Italiano of Buenos Aires, between November-December 2018. Patients of both sexes are included who carry out a protein-controlled diet from early diagnosis during the first year of life, until the moment of evaluation. Patients should have clinical evaluations with the team, through biochemical analysis and dietary records while taking the formula. The indexes and measurements taken were weight, size, size/age and weight/age or BMI index. They were categorized by sex and age. RESULTS: A group of 18 patients was included (56% female), within a range of 11 months - 24 years (with a median of 11,0 years). The diagnosis were 7 (39%) ornithine transcarbamylase deficiency, 6 (33%) phenylketonuria, 2 (13%) methylmalonic acidemia, 1 (6%) propionic acidemia, 1 (6%) citrullinemia type 1, and 1 (6%) maple syrup urine disease. In the evaluation of the weight/age or BMI/age n=1 (6%) presented underweight, n=12 (67%) normal weight, n=4 (22%) overweight and n=1 obesity. In the evaluation of size/age n=3 (17%) presented short stature, 66% of this patients had a diagnosis of organic acidemia. CONCLUSION AND DISCUSSION: The nutritional state was generally normal in the evaluated patients. The cases of short stature were associated to poor adherence to treatment, primarily by a failure to take the formula and/or wrong application of the protein-controlled diet. Hence, protein-controlled diet in these pathologies would have no negative impact on the nutritional state of the patients.