O-007 - GROWTH PATTERNS OF PKU PATIENTS IN THE FIRST YEARS OF LIFE

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INTRODUCTION: The first years of life are characterized by the highest growth rate of human life, and nutritional deficits could have a permanent impact on development. The aim of this study was to investigate growth patterns in the first 3 years of life of patients with phenylketonuria (PKU) and its association with metabolic control. ***METHODS:*** A retrospective cohort study was conducted in Hospital de Clínicas de Porto Alegre, Brazil. Patients with PKU diagnosed through NBS that started dietary treatment <6 months of age were included. PKU type was classified as classical PKU, mild or not determined. WHO Z-scores of weight-for-age (W/A), height-for-age (H/A), BMI-for-age (BMI/A) and head circumference-for-age (HC/A) were assessed at birth, before treatment, and at 6, 12, 18, 24 and 36 months of age. Mean and SD of phenylalanine (Phe) and tyrosine plasma levels were collected in year 1, 2 and 3. ***RESULTS:*** Forty-seven PKU patients were included (55% male), being 43% classical PKU, 48% mild, and 9% not determined. Pre-treatment measurements were similar among PKU types. Median age of the beginning of treatment was 45 days (range: 11-156). Mean Phe plasma levels were: 5.9±2.4 mg/dL in year 1; 6.4±2.8 mg/dL in year 2; and 7.5±3.8 mg/dL in year 3. In classical PKU patients, mean Phe plasma levels and Phe variability (measured by SD) were higher ($p<0.05$). Anthropometric indices W/A, H/A and BMI/A were within reference ranges for most patients, with no differences according PKU types. At 36 months, 1 patient presented short stature and 7 (15%) were overweight. An increase in overweight rate was observed in the second year of life and then remained stable. HC/A was within reference range for all but one patient (Z-score -2.3). However, classical PKU patients presented lower HC/A Z-scores at 6, 12 and 36 months (median at 36 months: -1.06). Phe variability, but not mean Phe, was inversely correlated with HC/A Z-scores at all points ($P<0.05$). ***CONCLUSIONS:*** Growth parameters were adequate for most patients, but the increase in overweight prevalence warrants attention. Our data suggest that avoid high Phe variability is important, since it was consistently associated with lower head circumference.