P-070

Variability of Phenylalanine concentrations over 24 hours using two different protein substiutes and changing dietary phenylalanine intake.

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Background: In PKU, there is evidence that glycomacropeptide (GMP-AA) may improve protein utilisation, which may stabilise blood phenylalanine (Phe) variability. In children with PKU, we studied the impact of GMP-AA on 24 hour blood Phe variability. A 6 week randomised controlled crossover study assessing blood Phe over 24 hours in children with PKU under three controlled dietary regimens, using 2 different protein substitutes CGMP-AA and Phe-free amino acids (L-AA).
Methods: We recruited 18 children with PKU, median age 10y (6–16), 7 boys. Each subject was randomised to 3 different regimens(R): R1, CGMP-AA and usual dietary Phe (CGMP+Phe); R2, CGMP-AA minus Phe from diet (CGMP minus Phe); R3, L-AA and usual Phe intake. Each regimen was for 14 days; on the last 2 days, Phe blood spots were collected 4hrly for 48 hr (08h,12h,16h,20h,24h,04h). Isocaloric intake was maintained.
Results: There was a consistent significant difference in median Phe concentrations over 24 h between each group: median Phe μmol/L; R1 v R2: 290 (30–580), 220(10–670) $p< 0.0001$; R1 v R3: 290 (30–580); 165 (10–640) $p< 0.0001$; R2 v R3: 220(10–670), 165 (10–640) $p=0.0009$. There was a significant difference in median Phe at each time point between R1vR2, $p=0.0027$ and R1vR3 $p< 0.0001$, but not between each time point for R2 v R3. Two groups show a reduction in Phe over time R2 = −1.67 Phe/hour and R3 = −2.05 Phe/hour, and R1 an increase over time+1.32 Phe/hour. Differences in the rate of change do not reach statistical significance. Of the 18 patients, 8 in the R1 group (44%) maintained target Phe levels for the study period, 10 in the R2 group (56%) and 7 in the R3 group (39%) [p = 0.052 – Chi-Square test]
Discussion: The residual Phe in CGMP-AA increases blood Phe control in children. Difference in the rate of change of Phe does not reach statistical significance although there is some evidence of GMP minus Phe giving the most consistent profile.
Conflict of Interest declared.