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Nutritional status in children with PKU: glycomacropeptide compared to phenylalanine free AA

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Background: In children with PKU, nutritional status is an important consideration particularly, with the primary nutrient supply being provided by protein substitutes (PS), supplemented with vitamins and minerals. Glycomacropeptide (CGMP-AA) has many suggested biological advantages. It is possible that it may enhance vitamin and mineral status, but no studies have compared the efficacy of CGMP-AA compared with conventional amino acid substitutes (L-AA) on nutritional status in children with PKU.

Methods: 48 children with PKU, median age 9.2y (5-16y), 28 boys, were divided into two groups; CGMP-AA (n=29) or L-AA (n=19)..A fasting morning venous blood sample was taken at baseline (when all the children were on L-AA) and after 6m intervention, and analysed for: zinc, selenium (plasma and whole blood), calcium, magnesium, phosphate, C-reactive protein, haemoglobin, mean cell volume, ferritin, vitamin B12 and 25 hydroxy vitamin D.

Results: At the end of 6m, median % of PS provided by CGMP-AA was 75% and L-AA, 25%. Whole blood and plasma selenium significantly increased from baseline to 6m (p=0.0002, p=0.0007) in the CGMP-AA compared to L-AA group. Within the CGMP-AA group from baseline to 6m whole blood selenium (p=< 0.0001) and plasma selenium (0.0005) significantly increased, but ferritin decreased (p=0.0006), but median values all remained within reference ranges (RR). Comparable changes were not observed in the L-AA group. All other nutritional parameters measured at baseline and 6m were within the RR. One exception in both groups was vitamin B12, which was above the RR at 6m. No differences were observed at baseline between groups.

Discussion: Both protein substitutes are efficacious in providing adequate nutritional status. There was a marked significant increase in plasma and whole blood selenium with CGMP-AA. It is possible the bioactive antioxidant properties of CGMP-AA and absorption in the gut may have a selenium sparing effect compared to a non-peptide-based feed.

Conflict of Interest declared.