

## GMP Focus - 2020-2021 in review

In 2020 and 2021 we continue to see lots of research published on the use of glycomacropeptide (GMP) in phenylketonuria (PKU).

### GMP in PKU - General

The PKU dietary handbook was published last year, with the aim of accompanying the 2017 European PKU guidelines. The European PKU guidelines stated that further evidence was required before recommendations could be given on the use of GMP in PKU. Three years on and the handbook can now provide some guidance;

- No significant increase in blood phenylalanine has so far been observed in adult patients using GMP (excluding maternal PKU patients).
- Care should be taken when introducing GMP in children with well-controlled PKU.
- Children with higher natural protein tolerance, due to a milder phenotype or through sapropterin usage, should tolerate the additional phenylalanine in GMP.

MacDonald *et al.* *Orphanet Journal of Rare Diseases* (2020) 15:171  
<https://doi.org/10.1186/s13023-020-01391-y>

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### PKU dietary handbook to accompany PKU guidelines



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## GMP in PKU - Children and teenagers

This study compared the post-prandial amino acid concentrations of three groups: The first group taking an amino acid-based (AA) protein substitute and the other two taking slightly different formulations of a GMP-based (GMP-AA) protein substitute, GMP-AA1 and GMP-AA2 (PKU sphere®). Two hours post-prandial amino acid blood profiles were taken for each group following a breakfast with 20g protein equivalent. All three groups saw significant rises in blood amino acid levels with any inter-group differences in the levels of individual amino acids attributable to differences in the amino acid profiles of the three protein substitutes. This suggests that protein substitutes derived from GMP-AA are absorbed similarly to that of AA protein substitutes.

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### **Preliminary Investigation to Review If a Glycomacropeptide Compared to L-Amino Acid Protein Substitute Alters the Pre- and Postprandial Amino Acid Profile in Children with Phenylketonuria**

Anne Daly <sup>1,\*</sup>, Sharon Evans <sup>1</sup>, Alex Pinto <sup>1</sup>, Richard Jackson <sup>2</sup>, Catherine Ashmore <sup>1</sup>, Júlio César Rocha <sup>3,4</sup> and Anita MacDonald <sup>1</sup>

This three-year prospective study, compared the impact of AA protein substitutes and GMP-AA protein substitute (PKU sphere) on both body composition and growth in 48 children with PKU. A trend towards improved growth and body composition was observed in children exclusively consuming PKU sphere, as their sole source of protein substitute long-term, when compared to children exclusively consuming AA protein substitutes or a combination of the two product types.

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### **Growth and Body Composition in PKU Children— A Three-Year Prospective Study Comparing the Effects of L-Amino Acid to Glycomacropeptide Protein Substitutes**

Anne Daly <sup>1,\*</sup>, Wolfgang Högl <sup>2</sup>, Nicola Crabtree <sup>1</sup>, Nick Shaw <sup>1</sup>, Sharon Evans <sup>1</sup>, Alex Pinto <sup>1</sup>, Richard Jackson <sup>3</sup>, Boyd J. Strauss <sup>4,5</sup>, Gisela Wilcox <sup>4,6</sup>, Júlio C. Rocha <sup>7,8</sup>, Catherine Ashmore <sup>1</sup> and Anita MacDonald <sup>1</sup>

Gut absorption of protein is thought to modulate satiety; however, the influence of the protein source or individual amino acids is not completely understood. This three-year study found that taking PKU sphere long-term had no impact on energy intake, weight gain or BMI when compared to consuming AA protein substitutes. No indication was found to support the relationship between GMP and satiety.

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### **The Impact of the Use of Glycomacropeptide on Satiety and Dietary Intake in Phenylketonuria**

Anne Daly <sup>1,\*</sup>, Sharon Evans <sup>1</sup>, Alex Pinto <sup>1</sup>, Richard Jackson <sup>2</sup>, Catherine Ashmore <sup>1</sup>, Júlio César Rocha <sup>3,4</sup> and Anita MacDonald <sup>1</sup>

Over a 36-month period, this study looked at the longitudinal bone density, mass and geometry in children with PKU taking either AA or GMP-AA (PKU sphere) protein substitutes as their main protein source. This highly comprehensive assessment of bone health in children and teenagers with good metabolic control, found that long-term PKU sphere consumption supports normal bone growth consistent with AA protein substitute consumption.



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## A Three-Year Longitudinal Study Comparing Bone Mass, Density, and Geometry Measured by DXA, pQCT, and Bone Turnover Markers in Children with PKU Taking L-Amino Acid or Glycomacropeptide Protein Substitutes

Anne Daly <sup>1,\*</sup>, Wolfgang Högl <sup>2</sup>, Nicola Crabtree <sup>1</sup>, Nick Shaw <sup>1</sup>, Sharon Evans <sup>1</sup>, Alex Pinto <sup>1</sup>, Richard Jackson <sup>3</sup>, Catherine Ashmore <sup>1</sup>, Júlio C. Rocha <sup>4,5</sup>, Boyd J. Strauss <sup>6,7</sup>, Gisela Wilcox <sup>6,8</sup>, William D. Fraser <sup>9</sup>, Jonathan C. Y. Tang <sup>9,10</sup> and Anita MacDonald <sup>1</sup>

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## GMP in PKU - Adults

This retrospective study evaluated the long-term impact on nutritional status and metabolic control of GMP-AA protein substitutes in an adult patient cohort, with a mean age of 28 years. GMP-AA protein substitutes provided a mean of 66+/-31% (range 23-100) of total protein equivalent intake. No significant changes in blood phenylalanine levels were found and all biochemical nutritional markers remained unchanged.

Pena et al. *Orphanet J Rare Dis* (2021) 16:84  
<https://doi.org/10.1186/s13023-021-01721-8>

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## Continuous use of glycomacropeptide in the nutritional management of patients with phenylketonuria: a clinical perspective

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PKU sphere is a food for special medical purposes and must be used under medical supervision.

This document is intended for healthcare professionals only.