Mevalia | AMINO ACIDS

A balanced combination of GMP-based and single amino acid products in the dietary management of PKU.

Powered by MEVALIA PKU GMPOWER.

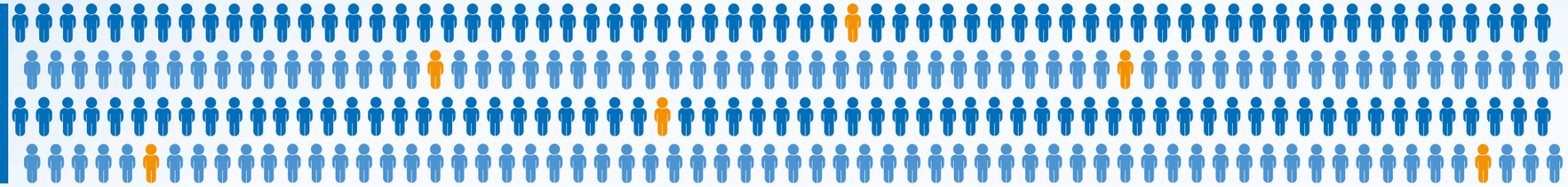
MEVALIA PKU GMPOWER is a product based on glycomacropeptide, which is naturally free from phenylalanine*. It is a preferred tasting protein supplement for the dietary management of PKU patients.

Endless innovation brings powerful solutions.

Dr Schär

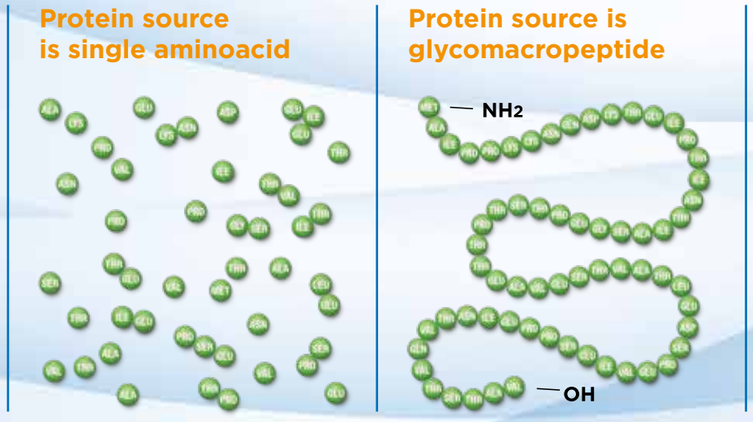
*The residual amount of Phe is due to the presence of minor amounts of other proteins/peptides [1].

Phenylketonuria (PKU) is an inborn error of amino acid (AA) metabolism due to mutations in phenylalanine hydroxylase gene. This causes decreased ability or inability to convert the phenylalanine (Phe) to tyrosine [1].



MEVALIA PKU GMPOWER

Phenylketonuria (PKU) is treated by a low-Phe diet, and the **major source** of protein equivalent is provided **by a Phe-free or low-Phe protein substitute,** in the form of either L-amino acids (L-AA) or casein glycomacropeptide (CGMP-AA).



Glycomacropeptide: an alternative to management of Phenylketonuria.
Tufail et all. International Journal of Scientific & Engineering Research Volume 8, Issue 12, December 2017

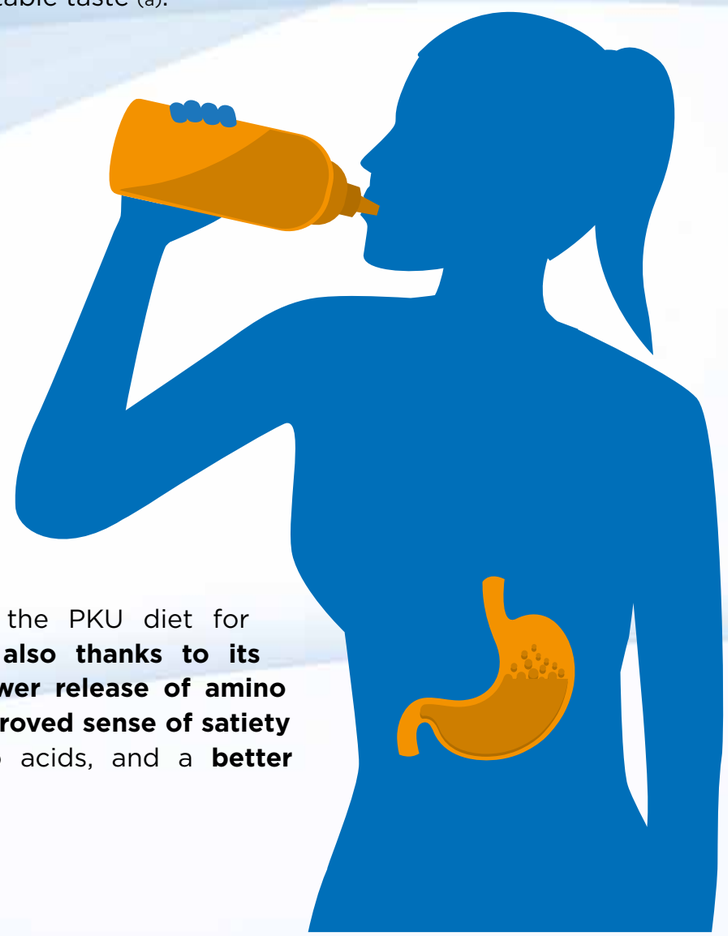
Glycomacropeptide (GMP) is a natural protein isolated from cheese whey, which is naturally free from phenylalanine. Recent studies showed that GMP is a valid alternative in conjunction with other treatment for patients with PKU who need phenylalanine [2] restriction, providing more satiety and allowing better adherence, especially in adolescence when compliance fails.

THE USE OF GMP IN THE PKU DIET (1)

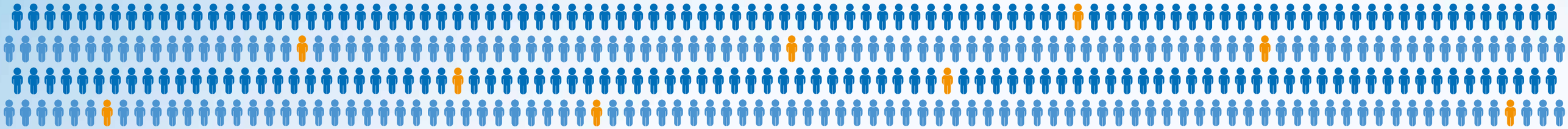
Due to a highly restrictive diet, and despite being paramount in PKU management, **compliance in PKU remains poor, especially during adolescence and adulthood [3].**

Typical issues linked to poor dietary compliance in PKU patients are **poor metabolic control and neurological outcomes.**

Glycomacropeptide has proven effective in promoting compliance, thanks to its palatable taste (a).

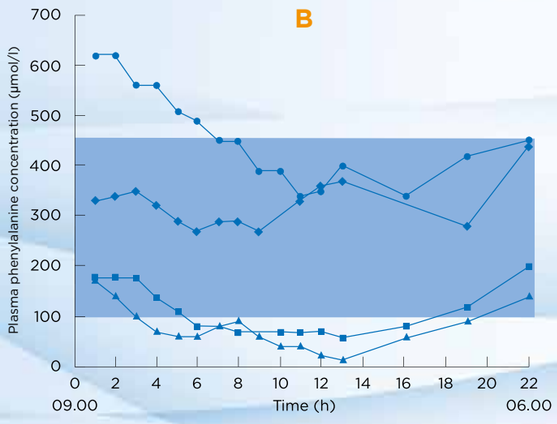


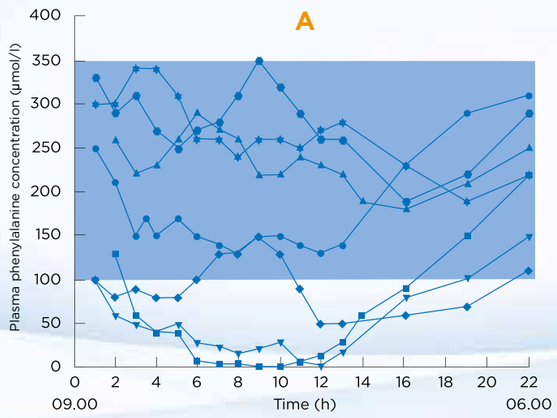
Not only is **GMP** useful in the PKU diet for supporting compliance, but **also thanks to its natural characteristics:** a **slower release of amino acids** in GMP results in an **improved sense of satiety** compared with single amino acids, and a **better utilization of protein (b).**

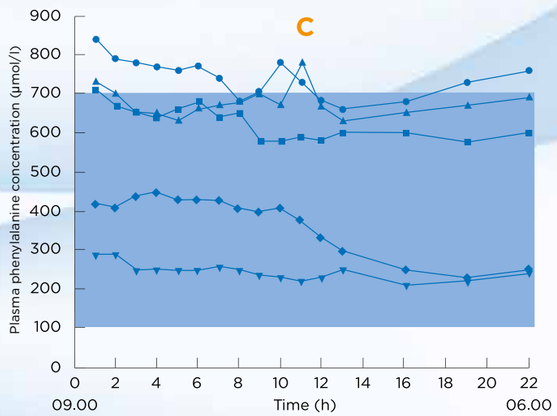


THE USE OF GMP IN THE PKU DIET (2)

Although a **higher Phe concentration in the blood** is sometimes recorded in PKU patients taking an **GMP-based** protein substitute (c), **a non-physiological 24 h blood Phe variability is also associated with Phe-free L-AA supplements [11; 12].**





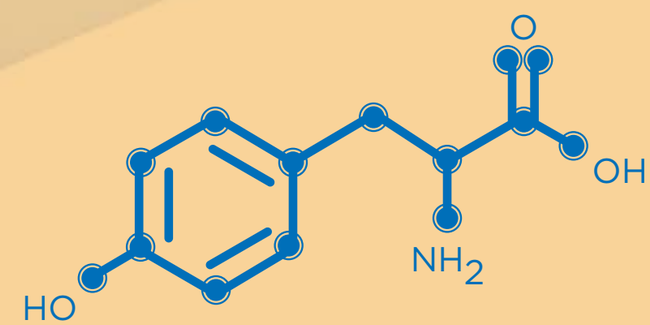


24 hour plasma phenylalanine concentrations in (A) 1–5 year olds (n = 7); (B) 6–9 year olds (n = 4); (C) 10–18 year olds (n = 5).
Does a single plasma phenylalanine predict quality of control in phenylketonuria? A MacDonald, G W Rylance, D Asplin, S K Hall, I W Booth

A review of the literature suggests that the cumulative effect of **day-to-day fluctuations over months and years** may affect cognition, **impacting on intelligence quotients [13,14].** A recent short study over 26 weeks by Feldmann concluded that Phe fluctuations were negatively correlated with IQ in children with mild PKU.
An **acceptable level of Phe in the blood also reduces the risk of hypophenylalaninemia,** which is linked to severe health issues [15].

Finally, **tyrosine is an important functional amino acid;** as a large neutral amino acid, **it competes at the blood–brain barrier with Phe.** There is also evidence that **Tyr from L-AA protein substitutes is less bioavailable than CGMP-AA protein substitutes, due to altered gut bacteria [16].** The **glycomacropeptide** structure appears to **lead to the stabilization of Phe concentrations,** with less fluctuation over a 24-h period compared to conventional amino acid formulas.

TYROSINE



a. Recent studies show that patients express a strong preference for GMP-based products rather than their usual AAs [4;5;6;7].
b. Van Calcar and colleagues [8] studied 11 subjects and reported lower Phe concentrations after an overnight fast using CGMP-AA compared to L-AA supplements, suggesting a slower release of AA in CGMP-AA. Other studies have also shown that protein synthesis and nitrogen retention are improved with 'complete' proteins compared to individual L-amino acids [9,10].
c. All median Phe levels were within European PKU target guidelines [17].

THE COMBINATION OF GMP AND SINGLE AMINO ACIDS

Studies have shown that in order to have the **highest benefit from the use of GMP-based protein supplements, phenylalanine intake** from diet should be reduced. This exchange should not be too problematic for the patient, thanks to the improved sense of satiety provided by the GMP compared to single amino acids.

The small and constant amount of phenylalanine taken in **through the GMP** product will, in turn, help to **avoid** phenylalanine fluctuations in the blood and **hypophenylalaninemia**.

The same balanced and efficient outcome in the management of PKU is proven with the **combination of 50% GMP-based products and 50% single amino acid products.**

This combination allows the **patient to control their phenylalanine intake, with positive effects in terms of sense of fullness, weight management, metabolic control and compliance, thanks to the pleasant taste and palatability of GMP [17].**

MEVALIA PKU GMPOWER (1)

MEVALIA PKU GMPOWER combines the benefits of a GMP-based product with a number of individual features. Enriched with essential fatty acids **DHA and EPA,** as well as in **GOS and FOS** prebiotics**, MEVALIA PKU GMPOWER is** one of the **most advanced solutions for the daily life of PKU patients.**

Essential fatty acids DHA and EPA

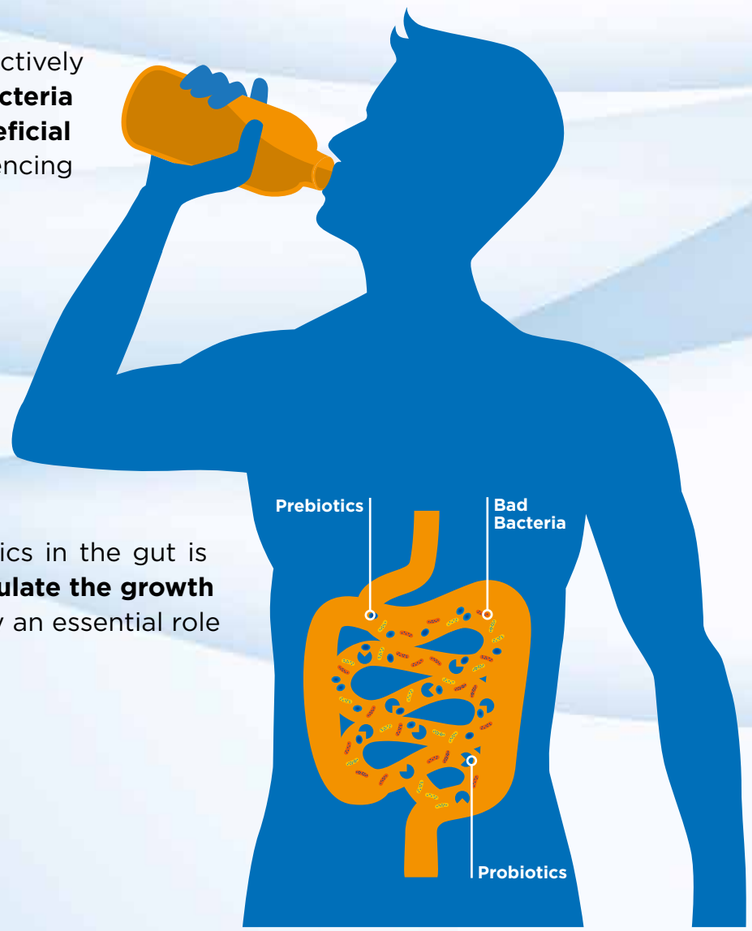
DHA and EPA are omega-3 fatty acids. These polyunsaturated fats play an important role in supporting health throughout life.
Evidence **suggests that children with PKU have reduced concentrations of DHA** in plasma and membrane phospholipids when compared to controls [18; 19; 20; 21].
We can conclude from these observations that a **supply of preformed n-3 LC-PUFA** is required to achieve **normal neural function in children with PKU [22]** in infancy and beyond [23].

MEVALIA PKU GMPOWER (2)

Prebiotics GOS and FOS

Considering the crucial **role of diet in shaping the gut microbiota,** i.e. the microbial community inhabiting the gastrointestinal tract [23], it is not surprising that **a low-protein PKU diet leads to microbial changes in phenylketonuric patients [24; 25].**

Prebiotics, by definition, selectively **increase certain** types of **bacteria** in the gut thought to be **beneficial to health** by positively influencing gut microbiota.



The fermentation of prebiotics in the gut is used by gut bacteria to **stimulate the growth of bifidobacteria,** which play an essential role in the **eubiosis condition.**

- Market research (d) shows that the majority of those interviewed prefer to take **MEVALIA PKU GMPOWER in the morning due to its mild flavor.**
- Its flexibility of use, thanks to the **convenience** of the mono-portion format, the possibility of **mixing MEVALIA PKU GMPOWER with different juices,** and its pleasant taste, judged as superior to single amino acid products, **help patients integrate the product in their daily routine.**
- Patients also report an **enhanced sense of fullness and better concentration. MEVALIA PKU GMPOWER** also seems to have **positive effects against cravings.**

MEVALIA PKU GMPOWER:

- ✓ **Contains a low ratio of PHE/TYR**
  14 mg of PHE and 1550 mg of TYR per 10 g PE
- ✓ **Is dilutable in low volume for better convenience**
  90 ml water per 10 g PE and 130 ml water per 20 g PE

d. 37 PKU people interviewed after taking GMPOWER at least once a day for a month. Market research carried out with the support of some metabolic centers in Austria and Germany, in the period from Jan-June 2019. Data on file.

CONCLUSIONS

The introduction of a GMP-based protein substitute in the PKU diet is easier and performs best when integrated with the intake of single amino acids.

A ratio of 50% GMP-based product to 50% single amino acid product permits patients to maintain low blood levels of phenylalanine, and to prevent phenylalanine fluctuations and hypophenylalaninemia.

This combination also requires no reduction in phenylalanine intake from low-protein foods [26].

References

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Nutrition declaration:		100 g	Per portion 23,4 g
Energy	KJ kcal	1352 319	316 75
Fat	g	1,7	0,4
of which saturates	g	0,7	0,2
Docosahexaenoic (DHA)	**mg**	**290**	**68**
Eicosapentaenoic acid (EPA)	**mg**	**64**	**15**
Carbohydrate	g	32	7,4
of which sugars	g	19	4,5
Fibre	g	3,0	0,7
of which GOS	**g**	**1,6**	**0,4**
of which FOS	**g**	**0,2**	**0,05**
Protein Equivalent	**g**	**43**	**10**
Salt	g	0,82	0,19
Amino Acids			
L-Alanine	g	2,76	0,65
L-Arginine	g	2,02	0,47
L-Aspartic Acid	g	4,80	1,12
L-Cystine	g	0,25	0,06
L-Histidine	g	1,28	0,30
L-Glutamic acid	g	4,17	0,98
L-Glutamine	g	2,76	0,65
Glycine	g	3,36	0,79
L-Isoleucine	g	1,84	0,43
L-Leucine	g	2,65	0,62
L-Lysine	g	3,47	0,81
L-Methionine	g	0,79	0,18
L-Phenylalanine	mg	61	14
L-Proline	g	5,39	1,26
L-Threonine	g	4,03	0,94
L-Tryptophan	g	0,77	0,18
L-Tyrosine	g	6,63	1,55
L-Valine	g	1,48	0,35

Nutrition declaration:		100 g	Per portion 23,4 g
L-Serine	g	1,42	0,33
Vitamins			
Vitamin A	µg	1248	292
Vitamin D	µg	37	8,75
Vitamin E	mg	25	5,85
Vitamin K	µg	75	18
Vitamin C	mg	187	44
Thiamin B1	mg	2,50	0,59
Riboflavin B2	mg	3,12	0,73
Niacin	mg	15	3,51
Vitamin B6	mg	2,50	0,59
Folic acid	µg	312	73
Vitamin B12	µg	5,00	1,17
Biotin	µg	75	18
Pantothenic Acid	mg	8,74	2,05
Minerals			
Sodium	mg	329	77
Potassium	mg	1753	410
Calcium	mg	1774	415
Phosporus	mg	1391	325
Magnesium	mg	329	77
Trace Elements			
Iron	mg	25	5,87
Zinc	mg	13	3,07
Copper	mg	1,31	0,31
Manganese	mg	1,31	0,31
Selenium	µg	62	15
Chromium	µg	50	12
Molybdenum	µg	73	17
Iodine	µg	276	65
Other Nutrients			
L-Carnitine	mg	25	5,86
Choline	mg	438	102
Inositol	mg	125	29

Osmolality: 825 mOsm/kg

1 sachet **90 ml water** **10 g protein**

12 g Mevalia PKU GMPOWER (3 measuring spoons) **45 ml water** **5 g protein**

MEVALIA PKU GMPOWER - IN BOX	
PACKAGING	ITEM NUMBER
Monoportion 10 g Protein 20 x 23,4 g	5653150700

MEVALIA PKU GMPOWER - IN TIN	
PACKAGING	ITEM NUMBER
468 g TIN	5653360701

MEVALIA PKU GMPOWER

- **PREFERRED TASTING***
- **Enriched with EPA and DHA (essential fatty acids) combined with GOS and FOS (prebiotics)**
- **With natural ingredients and a pleasant vanilla flavour**

*88% of patients who tasted Mevalia PKU GMPOWER during the ESPKU 2018 Congress rated the product as positive or very positive (ESPKU Congress 2018. Data on file).

MEVALIA AMINO ACIDS

2019/09

Dr. Schär develops preferred tasting amino acid mixtures, in liquid and powder form, for the dietary management of PKU patients.

- **Preferred tasting**
- **With natural ingredients and free from preservatives**
- **Convenience**

Foods for special medical purposes for use in the dietary management of phenylketonuria (PKU) and hyperphenylalaninemia (HPA) in children, adolescents and adults.

Mevalia | AMINO ACIDS

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