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# A case study outlining the transition from an amino acid based protein substitute to PKU sphere® in a thirteen year old.

Justin Ward, Specialist Paediatric Metabolic Dietitian, Bradford Teaching Hospitals.

## Patient Details & Medical History

**Age:**
13

**Gender:**

**Anthropometry at time of commencing PKU sphere:**
Weight (kg): 52.6 (< 91st centile) Height (cm): 151.3 (>50th centile)

**Biochemistry prior to commencing PKU sphere:**
Average Phenylalanine (Phe) 360-600µmol/l. Recent blood spot sampling, provided once monthly, had shown results at the upper end of the permitted range or above it. The patient associated this rise in Phe levels with a growing dislike of his Phe-free amino acid-based protein substitute (PFAA).

**Diet:**

Aiming for 1.5g/kg of total protein per day.
Dietary assessment revealed a variable daily protein intake of ≈ 60g protein equivalent (PE) (3 x 20g PE) from PFAA plus an estimated 20g of natural protein, from diet.
It is noteworthy that the patient does not strictly count 1g protein exchanges, but rather opts to avoid high protein foods such as meats, fish and dairy.

## Aim and Plan of Management

To transition from the PFAA to PKU sphere, a glycomacropeptide (GMP)-based protein substitute, in an attempt to reverse the gradual deterioration in metabolic control.

- To gradually introduce PKU sphere into the diet, 1 x 20g PE sachet at a time, with a corresponding decrease in PE from the PFAA.
- To increase Phe monitoring to once weekly and maintain a Phe level <600µmol/l during the transition phase.
- To investigate any Phe results >600µmol/l and assess whether the Phe inherent in PKU sphere (36mg/20gPE) was responsible.
- To ensure overall nutritional adequacy of the diet.
- To continue to monitor growth and development.

## Transition

Despite discussing the benefits of a staged transition from one product to the other, to allow monitoring of the impact of PKU sphere's Phe content, the patient opted to immediately switch all his PFAA to PKU sphere following PKU clinic. Furthermore, the patient continued to conduct blood spot sampling on a monthly basis as opposed to weekly which was recommended. Given these issues, the transition was instantaneous and difficult to monitor; nevertheless, a notable improvement in plasma Phe has been observed in the three results received since commencing PKU sphere (ranging between 113-364µmol/l).

## Outcome

A correlation was observed between the introduction of PKU sphere and an improvement in the patient's Phe control. Inevitably, there are limitations when drawing conclusions from this case report. Arguably, the most significant limitation is the lack of adherence to a fixed number of 1g protein exchanges during the pre and post transition period. The increased flexibility of self-selecting a low protein diet as opposed to following a formal regimen creates another variable that could influence overall Phe control. Nevertheless, the patient reported eating a diet similar diet pre and post transition, highlighting that a reduction in plasma Phe was achieved despite no quantifiable reduction in natural protein intake. In effect the patient has acted as his own control in a case-control study.

The relatively short time frame (3 months) between commencing PKU sphere and the production of this case study, in addition to the long distance between the patient's home and the clinic, hampered the retrieval of anthropometric data. As a consequence of this, any impact of PKU sphere on growth cannot be commented upon.

As mentioned previously, the patient did highlight that he struggled to consume the required amount of his previous PFAA due to the dislike of its taste, an issue which the GMP formula has remedied.

The patient has not reported any adverse effects whilst taking PKU sphere.

## Future Management for Similar Cases

From an academic perspective, this case does not represent the ideal transition from a PFAA to a GMP-based protein substitute. Future cases would ideally utilise patients who adhere to a strict and measured natural protein intake and who followed the approved protocol whilst integrating PKU sphere into the diet. Moreover, regular blood testing throughout the transition would highlight any deviation in plasma Phe levels in a timely manner and allow potential modification of the diet to ensure safety is maintained throughout the process.

Nonetheless, the outcome of this case study is overwhelmingly positive as our patient now has an enjoyable protein substitute that he takes in the prescribed amounts and is benefiting with much improved Phe control.


**References:**

1. Burgard P Lachmann, RH Walter J (2016) Inborn Metabolic Diseases: Diagnosis and Treatment. Edited by Jean-Marie Saudubray, Matthias R. Baumgartner and John H. Walter. Berlin: Springer

2. van Spronsen FJ, van Wegberg AM, Ahring K, et al (2017) Key European guidelines for the diagnosis and management of patients with phenylketonuria. The Lancet Diabetes & Endocrinology. DOI: http://dx.doi.org/10.1016/S2213-8587(16)30320-5.



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Vitaflo International Ltd, Suite 1.11, South Harrington Building,
182 Sefton Street, Brunswick Business Park, Liverpool L3 4BQ, UK.

Tel: 0151 709 9020
www.vitaflo.co.uk

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