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Introducing PKU sphere™ in an 8 year old with a low Phenylalanine (Phe) tolerance and managing Phe control.

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Patient Details & Medical History

Age: 8

Gender:

Weight: 24kg (25th percentile)

Height: 125cm (25th percentile)

Protein Equivalent (PE) from Amino Acid (AA) - based protein substitute: 60g/day (3 x 20g PE)

Natural protein intake: 4g/day **Total protein intake:** 2.7g/kg/day **Target phenylalanine (Phe) range:** 120-360µmol/L

Background

An 8-year-old girl taking conventional AA based protein substitute three times a day expressed an interest in taking a new Glycomacropeptide (GMP) - based protein substitute after tasting it in the PKU clinic. The median blood Phe concentration for her previous 12 months was 245 µmol/L with 76% of her blood spot Phe levels within the recommended target range for age. This was considered good control. Her growth, both weight and height were on the 25th centile, with an appropriate body mass index. She ate a limited variety of foods (low protein bread, potato products, salad and low protein biscuits only) despite many attempts to try to expand the variety of foods eaten in her diet. She was taking 60g/day PE from 3 x 20g PE AA based protein substitutes (2.5g protein/kg) and 4g/day of natural protein (200 mg/day Phe).

Introduction of (GMP - based protein substitute) PKU sphere20

The guidelines for the introduction of PKU sphere in section 2.5 of the 'Practical guide for PKU sphere' were followed. No adjustment or reduction of Phe exchanges was required.

Stage 1

1. One sachet of PKU sphere20 replaces one AA based protein substitute
2. Continue taking 2 x AA based protein substitutes
3. Weekly fasting capillary blood spots
4. Growth monitored frequently

Table 1. Introduction of 1 x PKU sphere20 (20g PE) in addition to 2 x AA based protein substitutes (40g PE)

Week	Phe µmol/L	Tyr µmol/L	Comments
1	240	40	No difficulties were observed and the new product was taken well. Weight stable.
2	190	50	
3	120	40	
4	280	50	

After 4 weeks of introducing one sachet of PKU sphere20, blood Phe concentration remained within target reference range.

Therefore, PKU sphere20 increased to 2 sachets per day with 1 x AA based protein substitute.

Stage 2

1. Two sachets of PKU sphere20 replace 2 x AA based protein substitutes
2. Continue taking 1 x 20g PE of AA based protein substitute
3. Weekly fasting capillary blood spots
4. Growth monitored frequently
5. Review after 4 weeks and change to all PKU sphere20 providing Phe levels are consistently below the target reference range

Table 2. Introduction of 2 x PKU sphere20 (40g PE) in addition to 1 x AA based protein substitute (20g PE)

Week	Phe µmol/L	Tyr µmol/L	Comments
5	260	30	No illness. Reported taking all protein substitute. Weight stable.
6	400	50	
7	120	40	
8	580	50	

Comments

There appeared to be no obvious reason for the increase in the blood Phe concentration. The patient had no illness and appeared to be adherent with her diet. However, her parents noticed she was reluctant to finish all her AA based protein substitute making total protein substitute intake less consistent than usual. A home visit established a few potential causes:

a) It was holiday time and the familiar daily routine of taking protein substitute was changed. Each morning she had a 'lie in' in bed therefore took the protein substitute later in the mornings.

b) Intake of food was less regular and Phe exchanges had not been measured accurately when eating outside the home.

c) She had attended a 'sleep-over' on two occasions and delayed taking her protein substitute till later in the morning.

Plan

As it appeared the fluctuating Phe control may have been either due to inadequate protein substitute intake or extra natural protein intake when eating out, it was agreed to repeat stage 2 and review after 4 weeks.

Table 3. Continuation of 2 x PKU sphere20 (40g PE) in addition to 1 x L-AA supplement (20g PE)

Week	Phe µmol/L	Tyr µmol/L	Comments
9	270	40	Routine established. Weight stable.
10	130	50	
11	190	40	
12	260	50	

Once her usual routine returned and she was back in school, the Phe concentrations returned to the patient's usual high standards. She was able to change to 3 sachets daily of PKU sphere20 after 8 weeks of following 2 sachets of PKU sphere20 and 1 pouch of AA PS.

Stage 3

1. Increase to three sachets of PKU sphere20
2. Stop AA protein substitute
3. Weekly fasting capillary blood spots
4. Growth monitored frequently

Table 4. Increase to 3 x PKU sphere20 (60g PE)

Week	Phe µmol/L	Tyr µmol/L	Comments
13	180	40	No illness. Reported taking all protein substitute. Weight stable.
14	220	50	
15	190	40	
16	240	40	

This young girl continued to have regular assessment of blood concentrations and she demonstrated good control of both Phe and Tyrosine concentrations (see Table 4.). Her growth continued to follow the 25th percentile for both height and weight and she continued to do well with her GMP-based protein substitute; PKU sphere20.

Key Messages

1. Transitioning to any new protein substitute needs regular monitoring.
2. If Phe control deteriorates it is important to consider changes to daily routine, in addition to other common causes of poor Phe control.
3. Using a methodical method of introducing a new dietary change provides confidence to the family and allows identification of the cause and the solution if any problems arise.

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All information correct at the time of print

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