

# Introducing PKU explore5, a second stage protein substitute and beginning complementary feeding.

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## Patient Details & Medical History

### Current Age:

14 months

### Gender:



### Diagnosis:

Phenylketonuria (PKU)

### Medical History

**Initial presentation:** 10-day old, breastfed infant referred to the metabolic service with a positive newborn screening result for Phenylketonuria.

**Neonatal screening result:** Phenylalanine (phe) 517  $\mu\text{mol/l}$  (normal - 60-90  $\mu\text{mol/l}$ ); Tyrosine 54  $\mu\text{mol/l}$  (normal - 30-120 $\mu\text{mol/l}$ )

**Confirmation result:** Phe 757  $\mu\text{mol/l}$ ; Tyrosine 54.7  $\mu\text{mol/l}$

### Relevant History

#### Anthropometry:

**Birth weight:** 3.0kg, weight on diagnosis: 2.9kg (9th centile), 2nd centile for length <sup>(1)</sup>

**Clinical/medication:** The infant was well at birth and breastfed with a top-up of expressed breast milk due to concerns of weight loss.

**Dietary:** On diagnosis, the infant commenced PKU start 20ml, 6 times per day prior to 6 breastfeeds, and then fed to appetite.

## Key Terms

**Phe exchange** - A measured amount of a protein containing food. A 50mg exchange system is used in the UK (1g of protein = 50mg phe = 1 phe exchange). Local practice on calculating phe exchanges varies internationally.

**Very Low Protein Foods** - Are low in protein or phenylalanine. They can be eaten without weighing or measuring.



## Dietetic Assessment

### Overall aim/goal:

To achieve target phe levels of 120-360  $\mu\text{mol/l}$  <sup>(2)</sup> during the introduction of both very low protein foods and the second stage protein substitute, PKU explore5.

To promote adequate growth and weight gain along centile charts.

### Estimated Daily Nutritional Requirements:

Total protein intake 3g/kg, 96 calories/kg <sup>(2)</sup>

### Dietetic Intervention:

Gradual, staged introduction of PKU explore5 at age ~6 months. Providing PKU explore5 ensured adequate protein equivalent (PE) intake for growth, whilst allowing appetite for very low protein foods and avoiding the need to increase the volume of PKU start to meet daily PE requirements.

## 6-8 months:

PKU explore5 was introduced from ~6 months of age prior to very low protein foods.

*This ensured that PKU explore 5 was tolerated and accepted before the introduction of very low protein foods.*

Half a sachet of PKU explore5 was made up according to instructions and one spoonful was given once daily (prior to PKU start and a breastfeed)

*This was at a time the infant was more likely to accept it.*

Once the infant was used to taking PKU explore5 from a spoon, the amount was increased gradually, until half a sachet of PKU explore5 was managed. There were no difficulties reported in the acceptance.

*Once the infant was accepting half a sachet of PKU explore5, PKU start was reduced by 125ml (2.5g PE).*

Once the child was happily taking half a sachet of PKU explore5, Mum was then advised to introduce very low protein foods after PKU explore5.

Very low protein foods such as butternut squash, sweet potato and carrots were among the first foods introduced. Mum offered these foods numerous times which helped aid acceptance. Gradually, the amount of very low protein foods the infant accepted increased at one meal, and then were introduced at a second mealtime.

*Starting with vegetables can help increase the likelihood of acceptance throughout infancy, which for the future may improve the variety of foods used in the restricted PKU diet <sup>(3,4)</sup>.*

As Mum started to reduce the number and frequency of breastfeeds these were replaced with phe exchanges such as potatoes, breakfast cereals and some vegetables such as peas, broccoli or cauliflower.


*Monitoring phe levels regularly can indicate a drop in intake of breastmilk so that phe containing foods can be used and increased.*

Blood spots were taken between once and twice a week to monitor his blood phe levels and they remained steady, within the target range of 120-360  $\mu\text{mol/l}$ .

Regular weight checks by the Health Visitor showed his weight had increased, and he continued to grow along the 9th centile for both weight and length.

## 8 months:

- **Weight:** 7.25kg (9th Centile)<sup>(1)</sup>
- **Protein Requirements:** 22g/day (3g/kg/day)<sup>(2)</sup>
- **Natural protein tolerance:** 9g/day
- **PKU start:** 180mls/day (3.6g PE/day)
- **PKU explore5:** 2 x 12.5g sachets/day (10g PE per day)

Meal Plan - 8 months old				Protein/PE (g)
<b>Breakfast</b>	<b>8g PKU explore5 powder</b>			3.3
	1 x 50mg phe exchange (breakfast cereal)			1
	Breastfeed			1
	40ml PKU start			0.8
<b>Mid-morning</b>	Breastfeed		1	
	30ml PKU start		0.6	
<b>Midday</b>	<b>8g PKU explore5 powder</b>			3.3
	Very low protein foods			
	Breastfeed			1
	40ml PKU start			0.8
<b>Mid-afternoon</b>	Breastfeed		1	
	30ml PKU start		0.6	
<b>Evening meal</b>	<b>8g PKU explore5 powder</b>			3.3
	Very low protein foods			
	Breastfeed			1
	40ml PKU start			0.8
<b>Bedtime/night-time</b>	Breastfeeds x 3		3	
<b>Total Protein</b>				<b>22.5g/day</b>

From 8 months until the infant's first birthday, the texture of foods became lumpier and finger foods such as low protein toast, pieces of fruit and cooked vegetables were offered regularly. He accepted a wide variety of very low protein foods, as well as specially manufactured low protein foods.

#### Ongoing Management:

- PKU explore5 was gradually increased to fully replace PKU start to meet protein equivalent needs by 14 months of age <sup>(5)</sup>.
- Natural protein needs are met by phe exchanges.
- Regular blood spots are taken to ensure blood phe levels are within target range.

## Result

### Monitoring/review:

Twice weekly blood spots ensured blood phe levels remained within target range.

Growth was monitored at the PKU clinic and by the child's Health Visitor - with weight increasing from the 9th to the 25th centile and length continuing along the 9th centile.

**Outcome measures:** Blood phe levels were within target range of 120-360  $\mu\text{mol/l}$  and a total daily protein intake of 3g/kg.

## Discussion

A reduction in PKU start was advised for this infant when both the family and dietitian were confident he was consistently taking half a sachet of PKU explore5 every day.

Gradually introducing and increasing PKU explore5 over several weeks gave the child's caregivers confidence with this transition and reduced the anxiety of quickly reducing PKU start.

Introducing PKU explore5 before starting complementary feeding sent a signal that PKU explore5 is essential and "non-negotiable". Getting this hurdle out of the way and then moving onto the complementary foods was beneficial in the long term.

The use of a food diary to record this infant's intake of PKU explore5, PKU start, very low protein foods and phe exchanges was vital for this family. It allowed them to keep track of how he was progressing, where he was up to in the transition and provided useful information for the dietitian to ensure that his fluid, energy, protein and micro nutrient needs were met.

### Key Learnings:

- Regular contact with the dietitian during this stage is essential so that alterations in amount and frequency of the PKU explore5 taken meets the infant's requirements.
- Introducing PKU explore5 prior to introducing very low protein foods may aid acceptance before other tastes are introduced.
- Regular monitoring of blood phe levels is needed alongside a food diary to ensure the transition from standard infant formula/breastfeeds to foods containing phe exchanges and PKU start to PKU explore5.
- Complementary feeding should begin once an infant shows appropriate behaviours.
- Ensuring multiple offerings of a variety of very low protein foods may help to increase their acceptance. This was seen in the varied diet of the child at 14 months.

## Conclusion

PKU explore5 was a successful choice of second stage protein substitute for this infant. His blood phe levels remained within target range, and he progressed well with solid foods and textures meeting feeding, growth and developmental milestones. Now, at 14 months, he is taking all of his protein equivalent as PKU explore5 and has a meal pattern that fits well with family life.

## References

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This information is intended for use by Healthcare Professionals only.

PKU explore5 is a Food for Special Medical Purposes and must be used under medical supervision. Suitable from 6 months of age. PKU explore5 is for the dietary management of phenylketonuria (PKU). Not suitable as a sole source of nutrition. For enteral use only. PKU explore5 contains **fish** (tuna). Not suitable for individuals with milk allergy.



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