

# Starting Solids and Introducing a Second Stage Protein Substitute (PKU gel™)

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## Introduction

Introducing solids to any infant's diet is a challenge for parents, with issues such as food refusal being commonly encountered. These issues are seen more frequently in infants with PKU. This case study highlights the process behind the successful introduction of solid foods to a young child with PKU, and the role a semi-solid protein substitute, PKU gel™, can play in this.

## Diagnosis

Alfie was diagnosed with PKU at the age of 10 days via newborn screening. Once diagnosed, Alfie was immediately prescribed a phenylalanine-free (phe-free) infant formula prior to every breastfeed. Alfie's control was excellent, and his parents took twice weekly blood samples to check his phe levels were in the target range (120 to 360  $\mu\text{mol/l}$ ).

### Key:

*Exchange-free* - Phenylalanine free or very low in phenylalanine foods.

*Exchange* - A measured amount of a protein containing food. In the UK, 50mg of phenylalanine = 1 phe exchange = 1g of natural protein

### References:

<sup>1</sup> [http://www.rcpch.ac.uk/system/files/protected/page/A4%20Boys%200-4YRS%20\(4th%20Jan%202013\).pdf](http://www.rcpch.ac.uk/system/files/protected/page/A4%20Boys%200-4YRS%20(4th%20Jan%202013).pdf) July 2017

<sup>2</sup> Medical Research Council. Recommendations on the dietary management of phenylketonuria. Archives of Disease in Children. 1993;68:426-7.

## 5 months - Starting Solids

Alfie showed signs of readiness for solid foods:

- Curious and interested when his sibling ate her meals
- Had good co-ordination – eyes and hands to mouth movements
- Could hold head up and sit well when supported

**Weight:** 7.5kg **Length:** 65.5cm - Both 50th centile<sup>1</sup>

**Total Protein Requirement:** 22.5g/day (3g/kg/day)<sup>2</sup>

### The dietitian advised Alfie's parents to:

Action	Rationale
Introduce low protein exchange-free vegetables and fruits into Alfie's diet (e.g. puréed sweet potato, carrot, or mango).	Introducing exchange-free vegetables/ fruits when the child is ready encourages acceptance of exchange-free foods that will not adversely affect blood phe levels.
Introduce exchange-free foods after phe-free infant formula and breast milk, starting with 1-2 teaspoons once a day.	Accustoms the infant to smooth/semi solid consistencies by 6 months of age in readiness for the second stage protein substitute.
Increase frequency and quantity of exchange-free foods until 8-12 teaspoons are taken at one time.	<p>This ensures the volume of phe-free infant formula and breast milk remains the same, so this will not adversely affect blood phe levels.</p> <p>Once this amount of exchange-free food is managed, it signifies readiness of the infant to progress taking natural protein containing foods (phe exchanges) that will replace the natural protein from breast milk.</p>

## Around 6 months - Introduction of PKU gel + exchanges

Breastfeeds were requested by Alfie's Mum to be gradually reduced in preparation for her return to work.

As Alfie's appetite for exchange-free foods increased, he was beginning to refuse his phe-free infant formula.

### The dietitian advised Alfie's parents to:

Action	Rationale
Introduce 5g PKU gel (2g PE) once per day at breakfast before exchange-free foods. An extra few mls of water can be added to PKU gel to make a thinner consistency if required.	PKU gel will begin to gradually replace the phe-free infant formula in a concentrated volume, so still leaving room for exchange-free solids, whilst ensuring phe-free protein needs are met.
Once 5g PKU gel (2g PE) is managed, adjust the volume of phe-free infant formula accordingly (100ml of phe-free infant formula = 2g PE).	PKU gel is given before exchange-free foods when the baby is hungry to encourage its adherence.
Offer water from a beaker after PKU gel is given.	Providing water in a beaker helps prevent any possible gastrointestinal effects from PKU gel as it is more concentrated than phe-free infant formula and a beaker helps the transition from a bottle
Before, or at the same time as introducing PKU gel, introduce measured amounts of protein containing foods (phe exchanges) by dropping 1 daytime breastfeed at breakfast and replace with equivalent phe containing food (1 phe exchange - 50 mg Phe) e.g. 8g of baby rice mixed with low protein milk and fruit puree. Give phe-free infant formula feed after meals.	Gradual introduction of PKU gel allows for volume of phe-free infant formula to be controlled and not exceed 600 mls/day in older infancy until it has been completely replaced by PKU gel by 12-15 months.  There is a window of opportunity at this time-once a taste for natural protein has been established, there is a risk of PKU gel non-acceptance.
Once PKU has been established at breakfast, introduce 5g PKU gel (2g PE) at the beginning of each mealtime over the next 2 weeks, ensuring water from a beaker is given after PKU gel	PKU gel will gradually replace the phe-free infant formula.  PKU gel before exchange-free foods and phe exchanges encourages adherence.
During these 2 weeks, also gradually drop another 2 daytime breastfeeds and replace each feed with phe containing food (1 exchange/50 mg Phe) e.g. broccoli (60g). Continue to give phe-free infant formula after meals.	Continues reduction of breastfeeds, and 1 phe exchange of food provides the equivalent amount of phe that was from breast milk.

## At 7 months

Alfie is now established on 5g PKU gel (2g PE) before breakfast, lunch and evening meal.

**Weight:** 8kg **Length:** 70cm - Both 50th centile<sup>1</sup>

**Total Protein Requirement:** 24g/day (3g/kg/day)<sup>2</sup>

	500ml Phe-free infant formula	PKU gel	Natural Protein
Current Intake	10g (PE)	6g (PE)	6g
Recommended intake to meet total protein requirement	10g (PE)	8g (PE) (Therefore, requires 2g additional protein equivalent from PKU gel)	6g

**Based on the information above, the dietitian advised Alfie's parents to:**

Action	Rationale
Increase PKU gel by 5g per day (2g PE). Provide 20g PKU gel (8g PE) before meals (in practical terms give 7g PKU gel x 3 daily) and a total of 500ml of phe-free infant formula daily over 3 feeds, followed by breastfeeds.	To ensure total daily protein needs are met.
Start to offer low protein finger foods e.g. soft permitted vegetable sticks, low protein toast "fingers"	Encourages self-feeding, helps to meet energy requirements, and promotes variety in the diet.

## 12 months onwards

At 13 months, Alfie was managing low protein meals and PKU gel well but refused his phe-free infant formula.

Mum was ready to stop breastfeeding completely.

**Weight:** 10kg **Length:** 77cm - Both 50th centile

**Total Protein Requirement:** 30g/day (3g/kg/day)<sup>2</sup>

	Phe-free infant formula	PKU gel	Natural Protein
Current Intake	0g	20g (PE)	6g
Recommended intake to meet total protein requirement	0g	24g (PE) (Therefore, requires 4g additional protein equivalent from PKU gel)	6g

**Based on the information above, the dietitian advised Alfie's parents to:**

Action	Rationale
Increase PKU gel by 10g per day (4g PE). Provide 60g PKU gel (20g PE) before meals (in practical terms give 20g PKU gel x 3 daily)	To completely replace the phe-free infant formula.
Increase phe exchanges from food.	To completely replace the phe from the dropped breastfeeds.
Give ProZero™ (protein-free alternative to milk) from a beaker cup during daytime and as a bedtime drink.	To provide calories to meet daily energy needs and as an alternative to milk.

## Challenges

Age	8 months	11 months	15 & 17 months
<b>What happened?</b>	Viral infection with raised temperature and irritability led to refusal of PKU gel, solid foods, phe-free infant formula and breastfeeds.	Teething, poor sleeping and irritable, with slightly raised phe levels. Teething meant Alfie couldn't tolerate a teat and gagged taking solid foods and PKU gel.	Teething with slightly raised temperature and phe levels led to refusing solid foods and PKU gel.
<b>Concerns</b>	Achieving daily nutritional needs was challenging. Giving PKU gel more difficult Impact on blood Phe control Parental anxiety		
<b>Solution/ Advice</b>	Give the same total amount of PKU gel; <ul style="list-style-type: none"> <li>• divide into 5 amounts per day rather than 3.</li> </ul> Maintain consistency: <ul style="list-style-type: none"> <li>• same person administering PKU gel,</li> <li>• use the same words of encouragement</li> <li>• prepare PKU gel in the same way each time.</li> </ul> Allow for PKU gel to take longer to administer Maintain in regular contact with parents		
<b>Outcome/ Result</b>	Gradual return to normal feeding over 7-14 days.		

## Outcome

Alfie is now 2 years old. He is a happy and healthy little boy who attends nursery 3 mornings a week. Alfie takes 3 sachets of PKU gel (30g PE) a day, before his breakfast, lunch and evening meal. He eats similar meals to his family, but using low protein foods. He tolerates 6g of natural protein per day (6 phe exchanges -each exchange is 50mg of phenylalanine).

## Key Messages

- Timing is crucial – there is a window of opportunity at around 6 months of age to introduce PKU gel. This will promote acceptance.
- PKU gel should be offered before or at the same time as natural protein/phe exchanges from food. This accustoms the infant to the taste of PKU gel before further dietary expansion.
- By 12-15 months of age, PKU gel should completely replace the phe-free infant formula.

PKU gel and ProZero are foods for special medical purposes and must be used under medical supervision.



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