



**explore™**

An introduction to complementary feeding and  
transitioning to a concentrated second stage protein  
substitute PKU explore™ in infants with phenylketonuria.



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Supporting education in the  
dietary management of rare diseases

## Disclaimer

This practical guide covers an introduction to complementary foods and transitioning to a concentrated, low volume spoonable protein substitute (**PKU explore**), in infants diagnosed with phenylketonuria (PKU).

It should be read in conjunction with local and national protocols. It is based on best practice over the last 20 years of introducing a concentrated spoonable protein substitute to infants with PKU from around six months of age<sup>(1)</sup>.

The guide **does not cover** the clinical management of PKU. Refer to section 4.1 for the international management guideline references.

This practical guide is:

- Only to be used by healthcare professionals.
- Not for use by patients or their families/caregivers.
- For guidance only and must not be used as a substitute for professional medical advice or management.

### Important notice

**PKU explore** is a food for special medical purposes (FSMP) for the dietary management of PKU (see section 5.3 for full **explore** range).

Use under medical supervision.

Not suitable for use as a sole source of nutrition.

Suitable from 6 months to 5 years of age.

For enteral use only.

**PKU explore** must only be consumed by individuals with proven PKU under supervision by the managing clinician or dietitian. Natural protein must be given in prescribed amounts to meet requirements.

**Introducing and adjusting PKU explore is based on the individual needs of the child. While practical examples are given in this guide, local practice may vary.**

**It is the responsibility of the managing healthcare professional to use their clinical judgment to introduce and adjust PKU explore in the most appropriate way for their individual patients.**

The product information contained in this guide, although accurate at the time of publication, is subject to change. To ensure accuracy, please refer to product labels or [www.vitafloweb.com](http://www.vitafloweb.com).

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## Key to symbols and terms used throughout this practical guide

Symbol	Term	Definition
	<b>Phe</b>	Phenylalanine
	<b>PKU</b>	Phenylketonuria
	<b>PE</b>	Protein Equivalent - the amount of protein provided by a protein substitute. Total protein intake = PE intake from protein substitute + natural protein intake.
		Breast milk/standard infant formula.
	<b>Phe-free formula for infants</b>	Phenylalanine free formula for infants.
	<b>PKU explore</b>	Concentrated spoonable protein substitute.
	<b>A measured amount of phe</b>	Throughout this guide, the term a <b>measured amount of phe</b> refers to a prescribed quantity of phe. This is mainly determined by target treatment blood phe concentrations. Systems for allocating phe amounts vary internationally. In the UK a 50 mg phe exchange system is used, 1g of natural protein = 50mg phe = 1 phe exchange. Other countries use allowance systems, where the individual counts the milligrams of the phe they consume.
	<b>Very low phe foods</b>	Throughout this guide, the term <b>very low phe foods</b> refers to foods that are very low in phenylalanine that could be incorporated into the diet without measurement (exchange-free). Where an allowance system is being used, every mg of phe may be counted as part of the patients daily allowance.



Practice on calculating **measured amounts of phe** and **very low phe food** intake may vary, adhere to local country / international guidelines.

When complementary foods and a second stage protein substitute are introduced to an infant with PKU, feeding can become more complex for both caregivers and healthcare professionals. These changes must not only support the child's developmental and nutritional needs but also maintain satisfactory growth and optimal metabolic control. New tastes are experienced through foods, and breast milk or standard infant formula are gradually replaced by measured amounts of phe containing solids. When choosing an appropriate protein substitute, it is always important to consider the age and developmental stage of the child.

One of the challenges of introducing complementary foods and a second stage protein substitute is to find a balance between giving a variety of very low protein foods and measured amounts of phe, and maintaining sufficient intake of the phe-free protein substitute to meet the infant's requirements. Historically, protein substitutes developed for young children were powders, designed to be reconstituted as liquids. When given to infants aged from 6 months, the high volume of liquid protein substitute may reduce appetite, delaying solid food progression. Administering the second stage protein substitute as a concentrated spoonable semi-solid, encourages appetite and therefore supports introduction of solid foods<sup>(1)</sup>.

Establishing the second stage protein substitute at the right time is essential to ensure it is accepted. Introducing **PKU explore** requires a gradual and progressive approach, increasing the amount according to individual needs. This enables the reduction of phe-free formula for infants, allowing appetite for foods. To support this process, caregivers will need education, ongoing support and practical guidance with clear instructions to help them with the challenges of complementary feeding and establishing the child onto a second stage protein substitute.

Good feeding practices for non-PKU infants should be followed when possible (see section 5.1). Differences occur with the types of first foods introduced and also the introduction of phe containing foods. To aid the acceptance of solid foods and support long term metabolic control, it is essential that this process is carefully managed<sup>(2)</sup>.

### Role of PKU explore

- **Provides a concentrated source of PE and micronutrients, which in the diet for PKU cannot be provided or are limited due to the restriction of protein rich foods.**
- **Ensures good feeding practice recommendations can be met, by reducing liquid intake and allowing appetite for solid foods to develop<sup>(3)</sup>.**
- **By helping meet infant feeding developmental milestones, PKU explore may reduce potential feeding problems and associated anxiety for both parents/caregivers and the child.**

VitaFlo has developed this practical guide for healthcare professionals on the use and introduction of **PKU explore** and complementary foods for infants at around 6 months of age with PKU. By following the suggested stepwise system for introducing **PKU explore**, it should help establish a suitable framework for protein substitute administration in infants and create a sound foundation for future years.

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## 1.1 What is PKU explore?

**PKU explore** is a concentrated powdered phe-free\* protein substitute, containing essential and non-essential amino acids, carbohydrate, sugar, vitamins, minerals, trace elements, arachidonic acid (ARA) and docosahexaenoic acid (DHA). It is intended for the dietary management of PKU from 6 months to 5 years of age.

**PKU explore** is reconstituted with a small amount of water to form a low volume smooth semi-solid paste that will hold consistency throughout mealtimes. Introducing **PKU explore** from around 6 months of age begins the transition from a phe-free formula for infants to the concentrated spoonable protein substitute. It is designed to help meet the infant's nutritional and developmental feeding needs; ensuring the goals of PKU dietary management are met, aiding growth and metabolic control.

Available in 2 pre-measured sachet size options.

Product Name	Premeasured sachet	Protein Equivalent (PE)	Flavour(s)	Age Indication
 <b>PKU explore5</b>	12.5g	5g	unflavoured	From 6 months - ideal for the first stages of complementary feeding
 <b>PKU explore10</b>	25g	10g	orange or raspberry	From 1 year - for when complementary feeding is more established and protein requirements increase with weight

**PKU explore5** follows on from phe-free formula for infants, easing the transition to the concentrated spoonable protein substitute. The flavoured options introduce new flavours as complementary feeding becomes more established and helps prevent taste fatigue.

Nutritional Information		PKU explore5 per 12.5g	PKU explore10 per 25g
Energy	kJ	181	351
	kcal	43	83
Fat	g	0.2	0.4
	of which saturates	g	0.17
ARA	mg	35	70
DHA	mg	18	35
Carbohydrate	g	5.3	9.8
	of which sugars	g	3.5
Protein equivalent	g	5.0	10
L-Phenylalanine*	g	-	-

**PKU explore** contains a wide range of micronutrients, for full nutritional information see [www.vitafloweb.com](http://www.vitafloweb.com)

\* No added phe. Phe may be present in trace amounts from other ingredients (<10mg/100g powder, <2mg/5g PE sachet, <4mg/10g PE sachet).

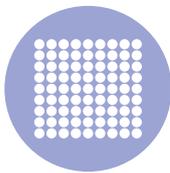
## 1.2 Nutritional features of PKU explore

### Protein Equivalent (PE)

Easy to calculate PE, supporting a staged transition from the phe-free formula for infants\* to the low volume **PKU explore**.

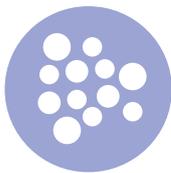


Volume of phe-free formula for infants	Protein Equivalent	Volume of PKU explore
 100ml	= 2g	=  5g PKU explore5 + 5ml cold water
 250ml	= 5g	=  1 sachet PKU explore5 (12.5g) + 12.5ml cold water
 500ml	= 10g	=  2 sachets PKU explore5 (25g) or 1 sachet PKU explore10 (25g) + 25ml cold water



### Micronutrients

A protein-restricted diet increases the risk of micronutrient deficiencies<sup>(4-6)</sup>. The composition of **PKU explore** contains a wide range of micronutrients including selenium, zinc, iron and calcium and a comprehensive range of vitamins including vitamin B12 and vitamin D important for nutrition and growth.



### Long Chain Polyunsaturated Fatty Acids (LCPUFAs)

LCPUFA intake is reduced as the rich food sources are restricted in a PKU diet. Children receiving a protein-restricted diet have shown deficiency in docosahexaenoic acid (DHA)<sup>(7,8)</sup>.

**PKU explore** meets requirements for DHA when  $\geq 30\text{g PE/day}$  is consumed for infants and young children<sup>(9)</sup>.

**PKU explore** has an ARA:DHA ratio of 2:1.

\* Based on a typical phe-free formula for infants.

## 2.1 Overview of the introduction of complementary feeding and PKU explore

The following chart shows an overview of the stepwise practical introduction of complementary feeding, both **very low phe foods** and **measured amounts of phe (exchanges)** + **PKU explore**, further details of each step follows in section 2.2.

**This is a guide only and every infant will progress at different rates.**

### Expected progression in feeding development



### Progressing from breast milk / standard infant formula to complementary foods

#### Breast milk / standard infant formula



Start to introduce tastes of **very low phe foods** first **AFTER** breast milk/standard infant formula + the phe-free formula for infants.

Gradually increase these foods in quantity, variety and then frequency.

Once accepting **very low phe foods** this signifies readiness to progress to introducing **PKU explore** and **measured amounts of phe**.

#### Introducing measured amounts of phe

Introduce a **measured amount of phe** to begin to gradually replace breast milk/standard infant formula.

Continue to increase the **measured amounts of phe** to individual tolerance and reduce breast milk/standard infant formula accordingly.

Top up meals with **very low phe foods** to appetite.

Continue to progress texture, introduce finger foods and encourage self feeding.

#### Total nutritional requirements are met through:

3 chopped/textured meals per day consisting of the allowance of

**measured amounts of phe** (to individual tolerance) +

**very low phe foods** to appetite + water

+

Divide **PKU explore** evenly over 3 meals.

Give **PKU explore** BEFORE food.

### Reducing phe-free formula for infants and introducing PKU explore

#### phe-free formula for infants



Do not decrease the volume of the phe-free formula for infants whilst introducing **very low phe food**.

#### Introducing PKU explore

Start with small amounts to taste.

As **PKU explore** is accepted increase the amount given, decrease the phe-free formula for infants accordingly.

**PKU explore** increases throughout the 2<sup>nd</sup> 6 months of life and beyond to meet growing protein needs.

Maintain volume of phe-free formula for infants at 500-600ml/day.

#### Continue transition

Gradually replace the phe-free formula for infants with **PKU explore**.



Encourage water throughout the day ensuring fluid requirements are met.



#### A phe-free formula for infants is important in the 1st year of life.



Although large volumes should be avoided to allow for developmental progression, continuing a certain amount helps contribute to ensure energy, fluid and other nutrient needs are met.

Appetite and oral intake of food increases throughout this period, which also enables energy requirements to be met.

#### Monitor

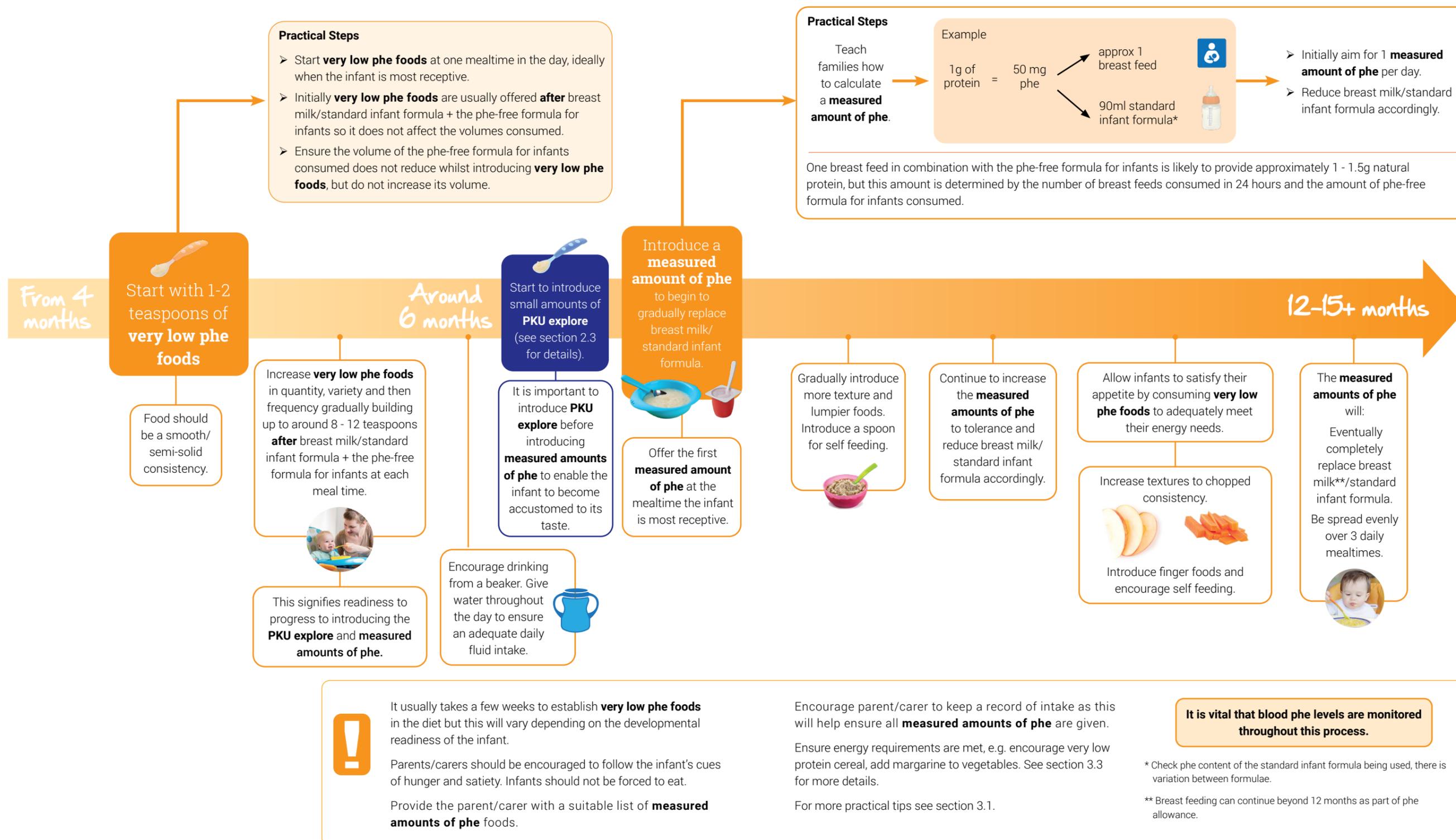
- Total protein equivalent
- Weight
- Fluid intake
- Stool pattern (ensure adequate fluid intake)

**It is vital that blood phe levels are monitored throughout this process.**

## 2.2 Stepwise introduction of complementary feeding

The following chart shows the stepwise practical introduction of complementary feeding; both **very low phe foods** and **measured amounts of phe**. In PKU introducing complementary food from 17-26 weeks of age can be advantageous as early exposure to food is likely to lead to acceptance of a wider range of **very low phe foods**, **measured amounts of phe** and **PKU explore**, whilst maintaining blood phe levels within treatment recommendations<sup>(2)</sup>.

When introducing food, it is best to give **very low phe foods** first as they do not adversely affect blood levels. This allows the infant to experience different tastes without affecting metabolic control. This **promotes a positive feeding experience** as there is less pressure to consume a required amount of food. Once accepting **very low phe foods** this signifies readiness to progress to introducing **PKU explore** and **measured amounts of phe (exchanges)**.



## 2.3 Stepwise introduction to PKU explore

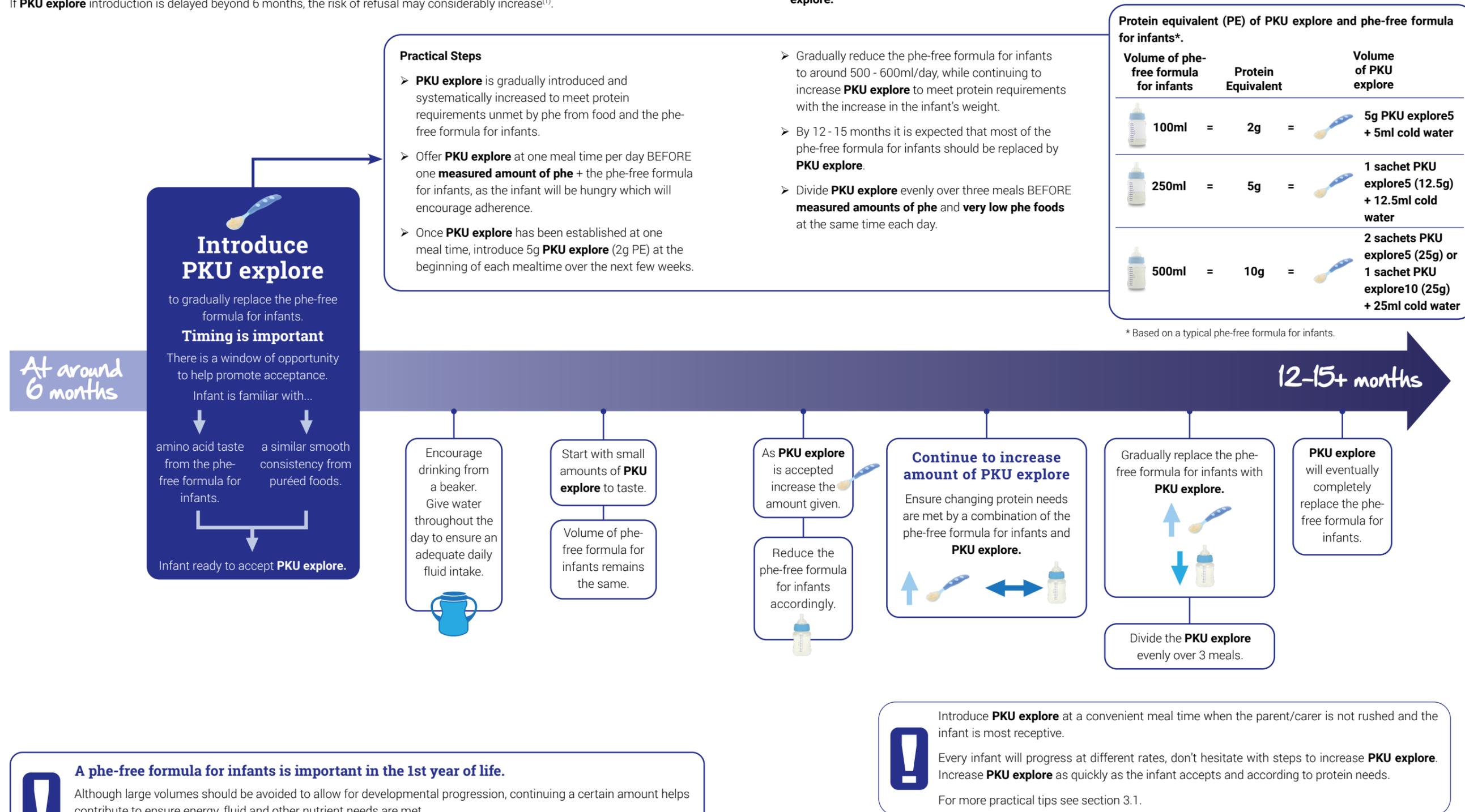
The following chart shows the stepwise introduction of PKU explore in more detail.

### Around 6 months - introduction to PKU explore

It is important to introduce **PKU explore** BEFORE introducing **measured amounts of phe (exchanges)** to enable the infant to become accustomed to its taste.

If **PKU explore** introduction is delayed beyond 6 months, the risk of refusal may considerably increase<sup>(1)</sup>.

Throughout this process calculate total protein requirements and consider phe from food. The remaining non-phe protein requirements are met by a combination of **PKU explore** and the phe-free formula for infants, until fully established on **PKU explore**.



At around 6 months

➔

Encourage drinking from a beaker. Give water throughout the day to ensure an adequate daily fluid intake.

Start with small amounts of PKU explore to taste.

Volume of phe-free formula for infants remains the same.

As PKU explore is accepted increase the amount given.

Reduce the phe-free formula for infants accordingly.

Continue to increase amount of PKU explore

Ensure changing protein needs are met by a combination of the phe-free formula for infants and PKU explore.

Gradually replace the phe-free formula for infants with PKU explore.

PKU explore will eventually completely replace the phe-free formula for infants.

Divide the PKU explore evenly over 3 meals.

Introduce PKU explore at a convenient meal time when the parent/carer is not rushed and the infant is most receptive.

**A phe-free formula for infants is important in the 1st year of life.**

Although large volumes should be avoided to allow for developmental progression, continuing a certain amount helps contribute to ensure energy, fluid and other nutrient needs are met. Appetite and oral intake of food increases throughout this period, which also enables energy requirements to be met.

**It is vital that blood phe levels are monitored throughout this process.**

The following table shows an example of how the infant's protein needs are met. It is **only a guide** and every infant will progress at different rates, it may therefore take longer to completely replace the phe-free formula for infants with **PKU explore**.

Weight	6kg	7kg	8kg	9kg	10kg	11kg	12kg
Approximate age	<b>Around 6 months</b>	→					<b>Around 24 months</b>
Phe-free formula for infants 	<b>700ml 14g PE</b>	<b>600ml 12g PE</b>	<b>600ml 12g PE</b>	<b>600ml 12g PE</b>	<b>400ml 8g PE</b>	<b>200ml 4g PE</b>	-
<b>PKU explore</b> 	Start with small amounts of <b>PKU explore</b> to taste.	<b>4g PKU explore x 3 = 5g PE</b>	<b>7g PKU explore x 3 = 8g PE</b>	<b>9g PKU explore x 3 = 11g PE</b>	<b>15g PKU explore x 3 = 18g PE</b>	<b>21g PKU explore x 3 = 25g PE</b>	<b>27g PKU explore x 3 = 32g PE</b>
<b>This infant tolerates 4g of natural protein*.</b> Breast milk/standard infant formula → <b>Measured amounts of phe food</b> 	<b>4g protein</b>	<b>4g protein</b>	<b>4g protein</b>	<b>4g protein</b>	<b>4g protein</b>	<b>4g protein</b>	<b>4g protein</b>
<b>Very low phe foods</b>	<b>Appetite and oral intake of food increases throughout this period, which enables energy requirements to be met.</b>						
<b>Total protein intake /day</b>	<b>18g</b>	<b>21g</b>	<b>24g</b>	<b>27g</b>	<b>30g</b>	<b>33g</b>	<b>36g</b>



The protein requirements in this example are calculated using the UK guideline of 3g total protein/kg/day<sup>(10, 11)</sup>, for infants and children 0 - 2 years old with PKU. Local recommendations and policies may differ on protein requirements for PKU infants and children and should be checked and adhered to where appropriate.

\* Phe tolerance will vary between individuals with PKU.

## 2.5 Sample meal plan - 10 month old boy

In the table below, a typical meal plan for meeting protein and energy requirements is shown:

<b>Weight of infant</b>	<b>10kg</b>
<b>Protein requirements</b>	<b>30g/day (3g/kg/day<sup>(10,11)</sup>)</b>
<b>Natural protein tolerance</b>	<b>4g</b>
<b>Phe-free formula for infants</b>	<b>550ml/day</b>
<b>PKU explore5</b>	<b>3 x sachets per day (37.5g powder)</b>



Remember to give a drink of water at each meal time.

			Protein/PE (g)	Energy (kcal)
<b>On waking</b>	200ml phe-free formula for infants		4	134
<b>Breakfast</b>	<b>1 sachet PKU explore5</b>		<b>5</b>	<b>43</b>
	10g Wheat biscuit (with protein free 'milk' alternative e.g. ProZero <sup>™**</sup> )		1	} 61
	<b>Very low phe foods</b> - low protein toast (half a slice)			
150ml phe-free formula for infants		3	102	
<b>Midday</b>	<b>1 sachet PKU explore5</b>		<b>5</b>	<b>43</b>
	40g mashed potato		0.5	} 100
	13g peas		0.5	
	<b>Very low phe foods</b> - small portion of low protein custard with chopped very low protein fruit			
<b>Evening meal</b>	<b>1 sachet PKU explore5</b>		<b>5</b>	<b>43</b>
	25g spinach		1	} 162
	<b>Very low phe foods</b> - low protein pasta, homemade vegetable sauce			
	20g yogurt <sup>†</sup>		1	
<b>Bedtime</b>	200ml phe-free formula for infants		4	134
	<b>Total</b>		<b>30g/day (3g/kg/day)</b>	<b>822kcal* (80kcal/kg/day)</b>

\* Calories estimated using manufacturer's labelling, food portion sizes<sup>(12)</sup> and food composition tables<sup>(13)</sup>.

\*\* **ProZero** is a protein-free alternative to milk suitable for use from 6 months of age.

† Check manufacturer's label.



If infant is still breast feeding, count as part of phe allowance.

Phe tolerance will vary between individuals with PKU.

### 3.1 Practical points for parents/caregivers for the successful introduction of PKU explore and complementary foods

Share practical advice with parents/caregivers to aid the successful introduction of **PKU explore** and complementary foods. Setting the right foundations and promoting good habits with a consistent approach from the beginning will support success in the long term.

#### Introducing PKU explore

**Be organised** - encourage parent/caregiver to be prepared, e.g. **PKU explore** powder pre-measured in a small bowl, baby bib, feeding spoon and water. A small 5ml syringe would be useful for measuring small amounts of water.

**Introducing PKU explore** - offer the infant **PKU explore** at a time of day when most receptive/interested and a convenient meal time when the parent/carer is not rushed.

**Routine** - give **PKU explore** at the same time and in the same way each day. Give at the start of each meal or half an hour before to ensure it is taken. Be consistent.

**Ensure all of the PKU explore is finished** - emphasise the importance of taking the prescribed amount.

**Same consistency** - prepare **PKU explore** to the **same consistency** every time. If this varies widely the infant may reject it due to unfamiliarity.

**Take it slowly and go at the infant's pace** - as with any new food it can take a little time for an infant to become used to **PKU explore**. **Persevere** with consistent timings and encouragement.

**Offer water** - to ensure an adequate daily fluid intake.

**Be patient** - it is common for any infant to spit out or refuse foods at times. It is no different with **PKU explore**, this is normal infant behaviour. Encourage parents to persevere and keep calm and continue with positive feeding techniques.

**During teething** - making **PKU explore** up with ice cold water may help with sore gums.

#### Introducing complementary foods

Encourage the use of **homemade foods** & provide information on suitable commercially available **very low phe foods**.

Provide families with lots of **practical advice** regarding cooking skills and ideas.

**Encourage batch cooking**, this is especially useful in the early stages when portion sizes are small.

**Repeat exposure** - remember it can take  $\geq 8$  times of offering the same food for an infant to become accustomed to the taste and accept it - **continue to offer foods even if at first refused**<sup>(14, 15)</sup>.

Encourage **eating with the family** and share some low protein meals/foods.

Establish **regular meal times**, but **avoid prolonged feeding**, keep to 20 - 30 minutes per meal.

Encourage parent/carer to keep a record of intake as this will help ensure all **measured amounts of phe** are given.

#### Provide information for other caregivers

Provide any new caregiver with an explanation of PKU.

Provide the caregiver with clear written instructions of the feeding plan and the necessary feeding equipment.

Emphasise the importance of ensuring the child finishes all of their **PKU explore** at each meal.

### Blood phe monitoring

Many factors can affect blood phe levels. Always check for causes of high or low blood phe level before making a change to feed regimen.

It is good practice to adjust the feed regimen after assessing more than one blood result, unless blood phe concentrations are very low or very high.

### Advice on illness

It is normal for an infant to refuse or have difficulties taking their usual amount of **PKU explore** when ill or teething. This may be an anxious time for parents/caregivers and so it is helpful to stay in regular contact with the family to give advice and reassurance and to assess how the infant is progressing. Sometimes parents/caregivers feel that switching protein substitute may help if the child is refusing their protein substitute. This should be avoided and the underlying issues of the illness or teething addressed.

It is very important that an infant continues to take their **PKU explore** during illness to help maintain metabolic control. However it may be necessary to recommend that the infant has the same daily dose, but administered in smaller more frequent amounts throughout the day, e.g. divide into five amounts per day rather than three. Allow time for **PKU explore** to take longer to administer.

**Continue to offer PKU explore throughout the day even when an infant refuses or is unwell. Giving an infant a 'day off' from their PKU protein substitute will adversely affect their metabolic control and give the wrong message to the infant and family. Stopping PKU explore, even for 24 hours, may create difficulties with its reintroduction.**

Extra fluids in the day may be necessary, particularly if the infant has vomiting or diarrhoea.

Parents should be given written advice on how to manage illness and should seek medical advice early.

### Feeding problems in toddlers

As with many toddlers, feeding problems are common in young children with PKU<sup>(16, 17)</sup>. Although children with PKU may be fussy for the same reasons as other children, some issues may be unique to these children, and place them at higher risk of developing feeding problems.

#### Strategies to minimise or improve feeding problems in an infant with PKU

- Good and consistent feeding routines are required from the commencement of complementary feeding and the introduction of **PKU explore**<sup>(1)</sup>.
- Parents/caregivers/families should eat together with the infant to promote a positive environment and social interaction where possible.
- Offer a suitable, healthy and varied low protein diet, but avoid offering too many food choices at one meal.
- Allow infants to satisfy their appetite by consuming **very low phe foods** to adequately meet but not exceed their energy needs. As these foods are **very low phe foods**, they will not adversely affect metabolic control.
- Limit excessive snacking.
- Encourage positive mealtime behaviours e.g. eat at the table together, allow toddler to help set the table.
- The infant should be seated during meals and distractions minimised.
- Encourage self-feeding at the appropriate stage.
- Give **PKU explore** in three equal doses before foods, at the same times each day.

### 3.3 Meeting energy requirements

When an infant transitions on to a solid diet, energy from breast milk or standard infant formula must be replaced by food. In a child with PKU, the transition from an phe-free formula for infants to a second stage protein substitute such as **PKU explore** creates an energy deficit to allow development of appetite for solid foods.

There has been some concern that the low energy density of a concentrated second stage protein substitute might not compensate for the energy content of the phe-free formula for infants and this might impact on growth and weight gain.

In an observational study<sup>(1)</sup> it was found that despite the lower energy content of the complementary feeding protein substitute compared to phe-free formula for infants, no infant was reported to have growth issues. It was assumed that an increase in solid food intake compensated for the energy discrepancy. This demonstrated that this type of protein substitute can be introduced without adverse effects on appetite or growth.

A later study looking at the growth, protein and energy intake in children with PKU taking a concentrated weaning protein substitute in the first 2 years of life showed that using the low volume protein substitute met phe-free protein requirements, facilitated transition to solid foods and supported normal growth<sup>(18)</sup>.

**It is important to make sure energy demands are met in this transitional period and beyond to facilitate growth and development.**

**General tips to increase energy intake if needed (section 3.4 for further details):**

- Fortify lower energy foods, such as vegetables and fruit, with extra energy, e.g. adding fat
- Offer more energy dense foods rather than increasing portion sizes
- Avoid drinks 1 hour before meals and offer after a meal so the child does not fill themselves up with fluid
- Have mealtimes when the infant is not too tired to encourage cooperation and appetite

**My First Low Protein Foods recipe book is available, which contains simple low protein recipes for the introduction of complementary foods.**

Some useful examples, of higher energy options include:

- Easy cheesie baked sweet potato
- Pancakes
- Cheeky cheesie bites
- Mini banana fritters



**PKU explore can be made up with ProZero instead of water to add extra calories. Make up PKU explore in the usual way but add ProZero instead of water. ProZero is a liquid blend of carbohydrate and fat designed as a protein free alternative to milk. It can be used in the dietary management of inborn errors of metabolism from 6 months of age.**

## 3.4 Tips on how to increase energy in the diet

The following methods can be used to increase the energy content of a low protein diet:

### Food sources

#### Low protein

Sweet potato  
Butternut squash  
Cassava  
Low protein pasta/rice/flour

#### Protein free

Gelatine free jellies  
Fruit purée  
Honey (after 1 year of age)

#### TIPS

These foods are easily incorporated into puddings or savoury dishes:

- Cassava to make a low protein crumble
- Low protein flours can be used to make cakes/pancakes and dough for bread sticks
- Sweet potato and butternut squash in mild curry sauces

#### TIPS

Sweet and sugary foods are easily incorporated into puddings or given as snacks:

- Add to low protein yogurt or fruit
- Add to low protein pancakes

Remember to advise to eat sugary foods within a meal and ensure dental hygiene.

#### Low protein

Low protein yogurt\*  
Mayonnaise\*  
Avocado  
Low protein cheese\*

#### Protein free

Cooking oils  
Margarine  
Butter  
Butter spreads  
Protein-free / low protein alternative to milk\*

#### TIPS

These foods are easily incorporated into puddings or savoury dishes:

- Mix cream\*/low protein yogurt with fruit to make fruit 'ice creams'
- Add suitable coconut milk\* to low protein vegetable sauces
- Mix cream\*/mayonnaise\* with avocado to make a spread for low protein toast

#### TIPS

Fats are easily incorporated into savoury meals:

- Lightly fry/roast vegetables
- Mix in to sweet potatoes/butternut squash
- Add into low protein tomato based sauces or mild curry sauces



**Some of these foods may be counted as part of the phe allowance, adhere to local country guidelines.**

**\* Check manufacturer's label for suitability.**

## 4.1 General references for the practical guide

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## 4.2 International management guideline references

### General reference

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## **5.0** Appendices

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**5.1 Complementary feeding an infant without PKU**

**5.2 References for complementary feeding an infant without PKU**

**5.3 explore range**

## 5.1 Complementary feeding an infant without PKU

This summary represents a guide to infant development and common practices for the introduction of complementary foods. Individual infants develop and progress at different rates. Advice should be tailored to the individual infant and nutritional requirements assessed on an individual basis. Complementary feeding practice varies internationally and this should be considered.

Developmental stage		Recommendations and practice	
<b>Stage 1</b> <b>Start</b> - Introduction of complementary foods begins not before 4 months but no later than 6 months <sup>(1-4)</sup>			
 <p>Infants begin to sit with assistance and develop some head control.</p> <p>Infants begin to mouth, hold and explore objects and may show interest in food.</p> <p>Infants may open their mouth in anticipation of a spoon.</p>	<p><b>Homemade or commercial complementary food</b></p> <p>Purée texture foods are introduced once per day when the infant is most receptive and progresses to 2-3 times per day according to acceptance.</p> <p>Complementary foods are offered before a milk feed.</p>	<p><b>Homemade or commercial complementary food</b></p> <p>Common first foods include baby rice, cereals, puréed fruit, puréed vegetables however all food groups, especially iron-rich sources<sup>(4,5)</sup> can be offered</p> <p>Foods to avoid include honey, added salt, added sugar<sup>(4)</sup>, rice drinks<sup>(6)</sup>, unpasteurised cheeses and raw shellfish.</p> <p>Foods may be refused initially and it can take more than 8-10 attempts before being accepted.</p>	<p><b>Breast feed/formula</b></p> <p>Milk feeds remain the same volume (around 120-150ml/kg/day) and complementary foods are experiential rather than nutritional at this stage.</p> <p>National policy should be consulted regarding recommendations for vitamin and mineral supplementation.</p>
<b>Stage 2</b> <b>Progress</b> - Progression of texture and variety of complementary foods			
 <p>The gag reflex to lumpy foods begins to decline.</p> <p>Infants develop skills to clear a spoon with their top lip.</p> <p>Mouth movements become more coordinated and chewing develops.</p>	<p><b>Homemade or commercial complementary food</b></p> <p>Frequency of complementary foods progresses to 3 small meals per day, offered before milk feeds.</p> <p>Food consistency can progress according to developmental stage from puréed to lumpy and mashed textures<sup>(1)</sup>.</p>	<p><b>Homemade or commercial complementary food</b></p> <p>Once 3 small meals are established, the variety of complementary foods widens to broaden acceptance and exposure.</p>	<p><b>Breast feed/formula</b></p> <p>A cup or beaker can be introduced from 6 months old.</p> <p>As the quantity of complementary foods increases, the volume of milk feeds reduce according to appetite but should remain at least 500-600mls/day<sup>(7)</sup>.</p> <p>Water is offered at mealtimes.</p>
<b>Stage 3</b> <b>Establish</b> - Further progression of texture and variety of complementary foods			
 <p>The presence of teeth allows infants to bite harder foods.</p> <p>Motor control develops to enable self-feeding from a spoon.</p>	<p><b>Homemade or commercial complementary food</b></p> <p>Infants progress to chopped foods, harder finger foods and foods that require more biting and chewing.</p> <p>Snacks are introduced between meals<sup>(3)</sup>.</p>	<p><b>Homemade or commercial complementary food</b></p> <p>Variety of foods progresses and is optimised to offer all food groups and nutrients.</p> <p>Mealtimes become more sociable.</p>	<p><b>Breast feed/formula</b></p> <p>Milk feeds gradually reduce to 2-3 times per day.</p> <p>Water is offered at mealtimes.</p>
<b>Stage 4</b> <b>Beyond the first year</b> - Joining in family meals and self feeding			
 <p>Infants start to recognise foods.</p> <p>Chewing is not fully mature but infants can cope with most textures.</p>	<p><b>Family meals</b></p> <p>Meals based on foods prepared for the rest of the family/caregivers are offered and social engagement at mealtimes is always encouraged.</p>	<p><b>Family meals</b></p> <p>Meals based on foods prepared for the rest of the family/caregivers are offered and social engagement at mealtimes is always encouraged.</p>	<p><b>Breast feed/cow's milk/follow-on formula</b></p> <p>Continue breast feeds according to mother's/infant's wishes.</p> <p>If formula fed, pasteurized whole cow's milk can replace formula if the complementary feeding diet is adequate to meet nutritional requirements<sup>(3)</sup> or follow-on formula can be used.</p> <p>Water is offered to meet fluid requirements.</p>

**Total nutritional requirements met by 3 chopped meals per day, snacks & drinks from a cup. Protein is achieved from a variety of food sources.**

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## 5.3 explore Range

**explore** is available for the following conditions, suitable from 6 months:

	Product / Condition	Precursor amino acid(s) free from	Flavour	Sachet Size	Protein Equivalent (PE)
	 <b>PKU explore5</b> Phenylketonuria (PKU)	Phenylalanine	unflavoured	12.5g	5g
	 <b>PKU explore10*</b> Phenylketonuria (PKU)	Phenylalanine	orange raspberry	25g	10g
	 <b>MSUD explore5</b> Maple Syrup Urine Disease (MSUD)	Valine Isoleucine Leucine	unflavoured	12.5g	5g
	 <b>HCU explore5</b> Homocystinuria (HCU)	Methionine	unflavoured	12.5g	5g
	 <b>TYR explore5</b> Tyrosinaemia (TYR)	Tyrosine Phenylalanine	unflavoured	12.5g	5g
	 <b>MMA/PA explore5</b> Methylmalonic Acidaemia/Propionic Acidaemia (MMA/PA)	Methionine Threonine Valine Low Isoleucine	unflavoured	12.5g	5g
	 <b>GA explore5</b> Glutaric Aciduria Type 1 (GA-I)	Lysine Low Tryptophan	unflavoured	12.5g	5g

It is essential that the correct **explore** product is prescribed for the particular condition. To make this easier the **explore** range has been given specific colours according to condition.

\* Suitable from 1 year of age.



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