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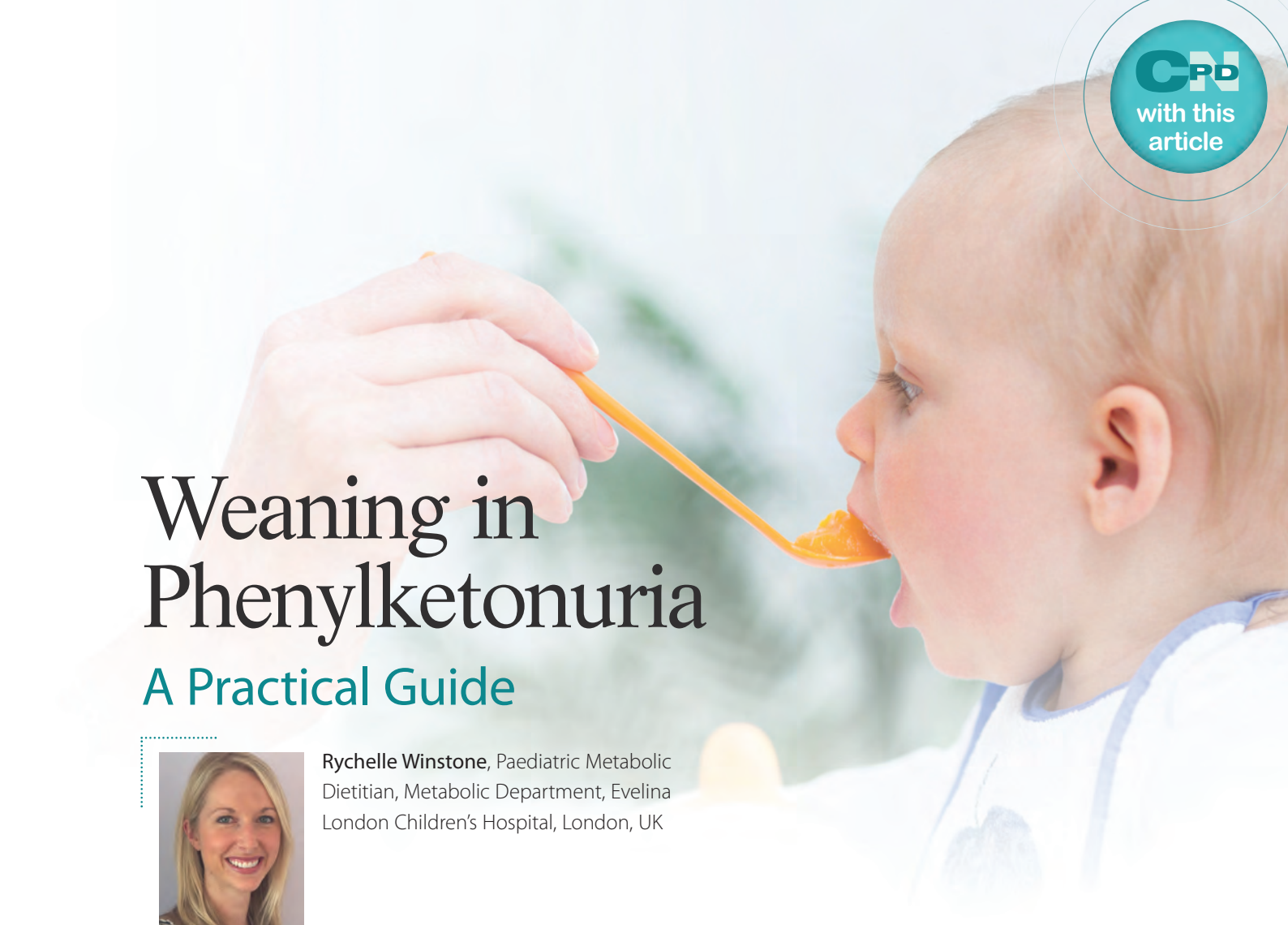


Weaning in Phenylketonuria

A Practical Guide

Rychelle Winstone, Paediatric Metabolic Dietitian, Metabolic Department, Evelina London Children's Hospital, London, UK

Phenylketonuria (PKU) is an inherited metabolic disease affecting approximately one in 10,000 babies born in the United Kingdom.[1] A deficiency in the enzyme phenylalanine hydroxylase (PAH) means that without early dietary treatment a build-up of the amino acid phenylalanine (phe) results in progressive and irreversible neurological impairment.[2] The cornerstone of dietary treatment is to restrict the intake of phe.[3]

This article will cover the introduction of complementary foods (weaning) for babies with PKU. It will give an insight into how our metabolic centre educates parents and caregivers to manage this important process. The considerations and challenges unique to this group will be discussed, and ways to support families to overcome them.

A baby with PKU will start life on a measured amount of phe provided by breastmilk or infant formula.[3] The extent that phe needs to be restricted will depend on several factors, such as the severity of PAH deficiency, age, weight, and growth rate.[3] Blood phe levels are closely monitored and phe intake is titrated against weekly blood spot test results.[2, 3] Measured phe in the diet is referred to as 'exchanges' (1 exchange = 50 mg of phe = approx. 1 g of natural protein).[3] Children with moderate to severe PKU typically tolerate around four to 10 exchanges per day.[3] A phe free amino acid supplement then makes up the shortfall in protein, calories and other nutrients that are needed for optimal growth and development.[2, 3] For babies this is in the form of a powdered or ready to feed formula. The total protein equivalent from exchanges and the phe free supplement should meet the recommended requirements for PKU (3 g/kg/d for children 0-2y[3]). As weaning progresses, foods naturally low in phe and low protein manufactured foods will also play key roles in the diet.

Getting started

Weaning is the transition that a baby makes from taking solely breast milk and/or infant formula, to including other foods and drinks,[4] and can have far reaching effects on eating behaviour and health.[5] In the UK, it is recommended that weaning is commenced at around six months of age (no later than 6 months and no earlier than 17 weeks).[4, 6] It has been suggested that babies weaned before six months of age (4-5 months) may accept a wider variety of new foods and have fewer feeding problems later on.[7, 8] Although, there is no formal consensus on the optimal weaning process for babies with PKU,[8] weaning should mirror the general recommendations wherever possible.[8]

Taking it in stages…

A common way of teaching weaning in PKU is dividing the process into stages. This way, parents and caregivers are given education in manageable chunks and are encouraged to enjoy each stage of weaning without worrying too much about what lies ahead. Reassurance is given during the first session that by the end of the four stage process they will be experts at feeding their baby a low phe diet! If possible, both caregivers should be present at each stage; information is also disseminated to others who regularly care for baby (e.g. grandparents, child minders). Interpreters are included if needed. See **Figure 1**.

Stage One: 'Getting Started'

During this first session, parents and caregivers are taught to introduce very low phe weaning foods. Breastmilk or infant formula continues to provide all exchanges, and phe free infant formula continues. Using very low phe solids has the distinct advantage of not needing to be measured or weighed; necessary when baby is still developing feeding skills.

A small list of very low phe ('free') fruits and vegetables is provided, e.g. apple, pear, sweet potato and parsnip. The National Society for Phenylketonuria (NSPKU) has analysed a wide variety of fruits and vegetables for their phe content, however, when it comes to manufactured baby foods that have not been analysed the protein content on the label needs to be used to approximate phe content.[3] This makes label reading very important, and it is a reoccurring theme right throughout the weaning stages. It gives parents and caregivers the ability to pick a product off the shelf and distinguish whether it is appropriate (a baby food is free if it contains <0.5 g/100 g protein[3]). One or two low protein prescription foods are also introduced at this stage, e.g. low protein rusks. Finally, information is given on aspartame: an artificial sweetener containing phe which must be avoided in food products and medicines.

Stage Two: 'Exploring Exchanges'

By the time we see families for Stage Two, baby's intake of weaning foods is becoming established and they are usually ready to start introducing exchanges with solid food. The goal of this session is to teach parents and caregivers the volume of breastmilk or infant formula that equates to an exchange, and then how to take this away when baby eats an exchange of solid food. The number of exchanges taken through solids is increased as baby and parents/caregivers are ready. A starter exchange list is given with a small selection of foods that can be included in homemade baby food (**Table One**). Label reading is revisited so that parents can calculate how much jarred or dried baby food corresponds to an exchange (see **Table Two** for an example). The goal is to spread exchanges as evenly throughout the day as possible.[2,3]

During Stage Two, 'Next Stage Supplement' is also mentioned. From around six months of age, growth and increasing protein requirements mean that the volume of phe free infant formula needed becomes unrealistic for an infant to take in addition to weaning foods. A more concentrated form of supplement needs to be introduced alongside the phe free infant formula, this is often in the form of a gel or paste and given with a spoon.

Stage Three: 'Learning more about Low Protein Foods'

At this stage baby is usually taking most, if not all, allocated exchanges with solids. We review how exchanges are being used and offer support and advice. A larger exchange list is given, and some more low protein prescription foods are introduced (e.g. bread, pasta, low protein milk substitute). Finger feeding is encouraged with free or exchanged finger foods (see **Table Three** for examples of free finger foods).

Stage Four: 'Filling in the Gaps'

This session is all about 'filling in the gaps' that have not been covered in the other stages. By now baby should be eating three meals of mashed or chopped food with suitable snacks in between, and formula intake has reduced to around 500 ml per day.[8] Label reading is covered again, this time to teach how to calculate exchanges per pack, slice, pot or biscuit (useful as baby's diet expands). Recipes and convenient supermarket products are featured, and booklets and pictorial lists are given out. The phe free supplement is likely to consist solely of a spoonable paste or gel, or an additional lower volume supplement drink may be taken in the morning and evening. See **Table Four**.

Figure 1: Staged Weaning Process in PKU

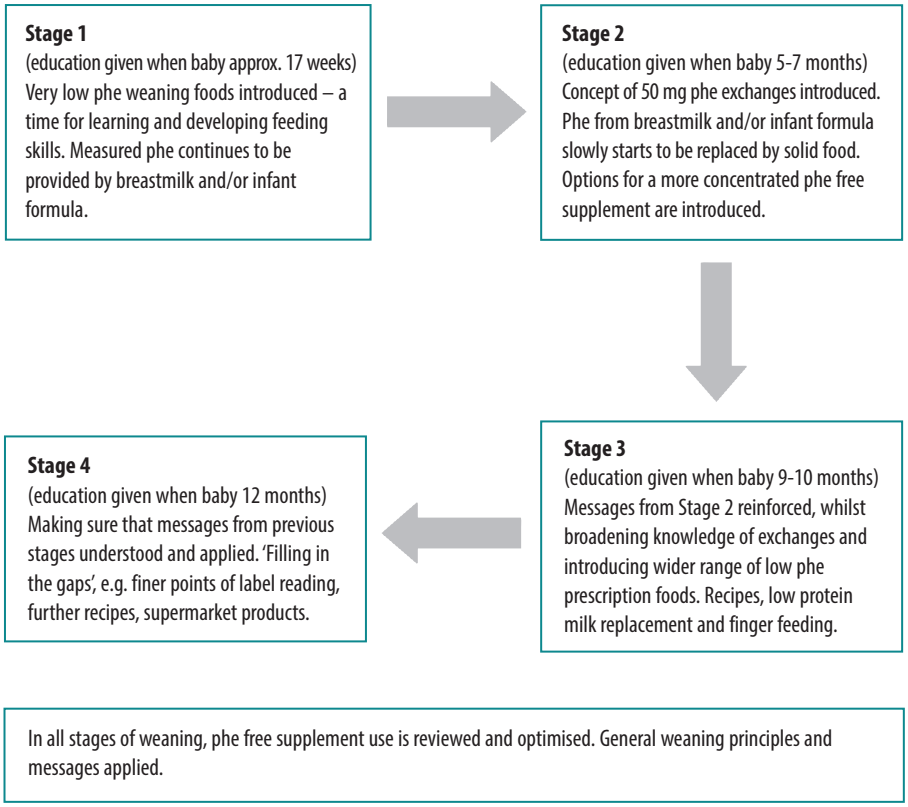


Table One: Starter Exchange List

The amount listed is equal to one exchange.

• Potato (mashed, boiled)	80 g
• Rice (boiled)	45 g
• Peas (fresh, frozen, boiled)	25 g
• Spinach (boiled)	25 g

Table Two: Example – Starting to Use Exchanges

To calculate an exchange of baby food = $\frac{100}{\text{(g protein per 100 g)}}$

E.g. Iron fortified baby rice (popular brand) has 7.8 g protein per 100 g powder

1 exchange (g) = 100/(g protein per 100 g)
= 100/7.8 = 13 g of baby rice

½ exchange = 7 g of baby rice

This should be made up with water or phe free formula. It can be mixed with pureed or mashed free fruits or vegetables (e.g. apple, or carrot) to give flavour and/or more volume.

Table Three: Examples of 'free' Finger Foods

- Lightly cooked free vegetables, e.g. carrot, courgette, sweet potato, parsnip
- Soft pieces of free fruit, e.g. pear, peach, kiwi, apricot, mango
- Low protein rusks
- Toasted fingers of low protein bread

Extra considerations

The prescriptive dietary treatment necessary in PKU brings with it an added focus on mealtimes and food, which can heighten parents' perceptions of feeding problems.[8] As parents move through the weaning process, a realisation that their baby's diet will be very different from the rest of the family often occurs and they can worry about future events such as birthday parties, nursery, school and family mealtimes. It has been shown that children with PKU can be more likely to be fed away from the rest of the family, and receive less positive prompting and reinforcement.[9] It is, therefore, very important to provide extra support at this early stage, and to help make the diet a normal part of baby's life.

Taking a phe free supplement at a prescribed amount, every day can potentially decrease appetite for solid foods.[8] The dietitian involved needs to be mindful of meeting baby's protein requirements, whilst ensuring that the substitute provided is in an achievable volume and a form that the baby can manage alongside weaning foods.[8] The appropriate recommendation of low protein prescription foods is similarly important. They should be included in baby's diet early on to ensure that they are readily accepted. As children grow, the number of exchanges does not change a great deal but the amount of food they need does. Many of the foods available for prescription in the UK can be high in sugar and/or salt and are not suitable for young children. Parents and caregivers should be advised on appropriate use by their dietitian.

Challenges and overcoming them

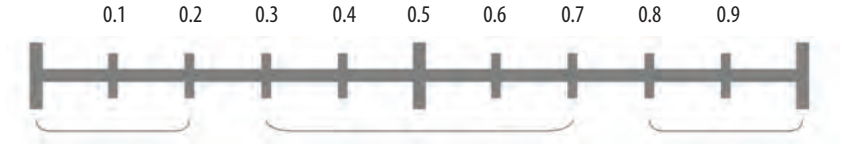
One of the greatest challenges families face is organisation, which is so important for the diet to be successful. Often families need help to organise information received. Each weaning stage is colour coded, and families are given a folder with clear pockets so that after each stage they can add to their information pack. This folder also has space to record blood test results and recommended number of exchanges. Electronic scales and calculators are given out to assist with calculating and weighing foods, and using devices such as Smartphones is encouraged to remind families to take bloodspot tests.

Literacy and mathematical skills varies widely across families that we see. Limited education or learning difficulties can mean that reading food labels or using calculations is difficult or impossible. Alternatively, verbal and/or written English may be limited or non-existent. Extra time is always spent with these families, and individually tailored pictorial lists and simple meal plans are used. It is vital to never assume basic skills. For example, some of our families struggle to round decimal places up or down when calculating. **Figure 2** shows a simple resource that we have found very useful when teaching label reading.

Table Four: Typical Diet for a 12-month Old with PKU *(5 exchanges per day)*

On waking	Phe free formula drink	4 g PE
Breakfast	Phe free supplement – served with a spoon	5 g PE
	Exchange of breakfast cereal with low protein milk substitute	(1 exchange)
	Fingers of low protein toast with butter	(0 exchanges)
Snack	Apple slices and raisins	(0 exchanges)
Lunch	Phe free supplement – served with a spoon	5 g PE
	Exchange of pasta with an exchange of green peas	(2 exchanges)
	Low protein pasta added if needed to bulk meal	
	Tomato based pasta sauce (free ingredients only)	(0 exchanges)
	Slices of cucumber and red and yellow peppers	(0 exchanges)
Snack	Low protein biscuit or pieces of banana	(0 exchanges)
Dinner	Phe free supplement – served with a spoon	5 g PE
	Exchange of mashed potato	(1 exchange)
	Exchange of baked beans	(1 exchange)
	Cooked carrots and green beans	(0 exchanges)
	Custard made with low protein milk substitute with cooked apple	(0 exchanges)
Before bed	Phe free formula drink	4 g PE
Water or diluted fruit juice offered throughout the day in a beaker		(0 exchanges)
		Total protein equivalent = 28 g per day (3.1 g/kg/d) (weight = 9 kg)

Figure 2: Rounding Decimals in Food Labels



Round down to lower number
e.g. 0.2 g protein = 0 exchanges
1.1 g protein = 1 exchange

Round to ½ exchange
e.g. 0.4 g protein = ½ exchange
1.7 g protein = 1 ½ exchanges

Round up to next number
e.g. 0.9 g protein = 1 exchange
1.8 g protein = 2 exchanges

Many parents and caregivers that we see have limited cooking skills and find the prospect of preparing homemade weaning foods daunting. Explaining and demonstrating simple things, such as the basic equipment needed, how to boil vegetables, and how to freeze baby food in ice cube trays for later use can make all the difference. This can help set parents and caregivers up for preparing home cooked low protein meals as babies grow and their diet diversifies. For families where cooking from scratch is unrealistic, advising on appropriate ready-made options, and how to combine these into meals is essential. Talking to families about the food they eat at home is very important to be able to tailor dietary advice to culture and lifestyle, and make sure that baby is being fed similar style food to the rest of the family.

Summary

Weaning in PKU forms the knowledge base that is used for a lifelong therapeutic diet. Dietitians guiding families through this process should apply normal weaning principles where possible, while being mindful of the extra considerations unique to the condition and to individual families. Advice should be provided by a metabolic team which includes a paediatric dietitian experienced in the care of children with PKU.[8]

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