PKU start™

A practical guide to the introduction and use of **PKU start**™, a phenylalanine-free formula.

Vitaflo in Association With You

Supporting education in the dietary management of rare diseases

Disclaimer

This practical guide is for the use of **PKU start** in the dietary management of an infant diagnosed with phenylketonuria (PKU). It should be utilised in conjunction with local and national protocols.

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

The product information presented in this guide, although correct at the date of publication, is subject to change. To ensure accuracy, please refer to product labels.

Important notice

PKU start is a food for special medical purposes (FSMP) for the dietary management of phenylketonuria (PKU).

Use under medical supervision.

Not suitable for use as a sole source of nutrition.

Suitable from birth.

For enteral use only.

PKU start must only be consumed by individuals with proven PKU in conjunction with breast milk or standard infant formula to provide the phenylalanine, fluid and general nutritional requirements of the infant in quantities as advised by a clinician or dietitian.

Introducing and adjusting PKU start is based on the individual, practical examples are given in this guide, however international and local practice may vary.

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

VIA (Vitaflo In Association) work in collaboration with healthcare professionals from around the globe to create product and disorder-specific support materials.

Collaborators

Vitaflo® dietitians in collaboration with Professor Anita MacDonald OBE, BSc, PhD, Consultant Dietitian, Birmingham Children's Hospital, UK.

Foreword

Phenylketonuria (PKU) is an autosomal recessive, inborn error of amino acid metabolism. Without treatment, most children develop profound and irreversible intellectual disability[1]. PKU is diagnosed at birth via new born screening. European and American guidelines strongly advocate that an infant with a phe level consistently ≥ 360 µmol/L is referred to a specialist metabolic centre to start dietary intervention no later than 10 days of life. It is recommended that a target range of 120 to 360 µmol/L is achieved within 2 weeks of life[2,3]. The feeding of infants with PKU is complex and multi factorial. It is important that a low phe diet supports optimal growth, development and mental functioning whilst meeting nutritional requirements.

If an infant with PKU solely received breast milk (BM) or standard infant formula (SIF), the phe intake would exceed their tolerance and result in increased plasma phe levels. However, as phe remains an essential amino acid, infants require a small, controlled amount of either BM or SIF to provide this phe to ensure adequate growth and development. In infants this is achieved by the restriction of phe via the introduction of a phe-free formula, in combination with BM/SIF[3].

Formula free from phe is critical in the dietary management of infants with PKU to prevent protein deficiency and optimise metabolic control. It should provide a balanced amino acid profile, except for phe, as well as providing carbohydrate, fat, vitamins, minerals and trace elements. For the majority of patients with PKU, the phe-free formula provides >75% of the total daily protein intake[4].

Clinical evaluation of PKU start

PKU start is a phe-free formula designed for infants with PKU. Three metabolic centres in the UK conducted a research study, to examine the efficacy, acceptance and tolerance of **PKU start**. Infants with PKU, aged from 5 weeks to 9.5 months, diagnosed by newborn screening were studied.

Findings showed that infants easily transferred onto PKU start, grew and gained weight satisfactorily with clinically acceptable blood phe control during the 28-day study[5].

This resource has been developed to offer practical guidance of how to introduce **PKU start** to a newly diagnosed infant with PKU, who is either being breast fed or fed with standard infant formula and a brief outline of progression on to weaning.

Prof. Anita MacDonald

PKU start

PKU start is an amino acid based powdered phenylalanine-free* formula containing other essential and non-essential amino acids, carbohydrate, fat, vitamins, minerals, trace elements, arachidonic acid (ARA) and docosahexanoic acid (DHA). **PKU start** provides all the essential nutrients required by an infant, except phenylalanine. It has been developed to comply with all relevant worldwide regulations for infant FSMPs. **PKU start** is designed to be used in the dietary management of PKU from birth.

* No added phenylalanine. Phenylalanine may be present in trace amounts from other ingredients (<10mg/100g; <1.5mg/100ml reconstituted product).

Key to symbols and abbreviations used throughout this guide.

Symbol	Abbreviation	Definition
	phe	phenylalanine
	**BM** **BF**	Breast milk Breast feed
	**SIF**	Standard infant formula
	**phe-free formula**	phenylalanine-free formula (**PKU start**)
		Concentrated second stage protein substitute (SSPS)

Throughout this guide we refer to measured portions of natural protein as phe exchanges (1g natural protein = 50mg of phe = 1 phe exchange).

Practice on calculating phe intakes varies between centres and internationally. Local procedures should be observed and adhered to when giving families/caregivers advice.

Contents

1.0 Features of PKU start

1.0 Features of PKU start

Up-to-date nutritional profile

Based on the latest scientific evidence on the nutritional content of infant formula[6] and recent research in PKU.

Complements Vitaflo's second stage protein substitute (SSPS) range.

The nutritional profile of **PKU start** supports transition to a SSPS.

2g PE = 100ml **PKU start** = 5g **PKU gel powder**

Well tolerated and accepted

Infants easily transferred onto **PKU start**, grew and gained weight satisfactorily with clinically acceptable blood phe control during the 28-day study[5].

Non-medical designed packaging - similar to SIF

Helps add normality to the diet at what can be an unsettling time for families/caregivers.

Mirrors preparation instructions of infant formulae.

PKU start is easy to prepare - familiar to healthcare professionals and families/ caregivers.

2.0 Application of PKU start

2.1 Overview of feeding an infant with PKU

All newly diagnosed infants with PKU are started on a phe-free formula immediately after diagnosis is confirmed. The aim is to achieve a rapid reduction in blood phe levels. Once plasma phe levels are below 1000µmol/l, a source of natural protein, either BM/SIF is reintroduced and given in combination with a phe-free formula.

Feeding an infant with PKU is a balance between providing enough phe and enough phe-free formula. Full term infants with PKU have a minimum phe requirement so are unlikely to need < 150mg of phe/day[7].

As with all new mums, breast feeding should be encouraged and supported with relevant healthcare professionals input as appropriate.

Breast fed infant

Phe-free formula combined with controlled amounts of BM is able to maintain satisfactory blood phe control and growth whilst still providing breast feeding benefits[2]. BM also has the added advantage of being low in phe - 46mg/100ml compared with SIF which is is on average approximately 60mg/100ml.

Breast feeding an infant with PKU is based on the principle of giving a measured volume of phe-free formula (**PKU start**) before each breast feed. This inhibits the infant's appetite and reduces suckling, thus decreasing the amount of breast milk consumed and therefore phe intake. Babies can still feed on demand, varying the quantity of feeds from day to day provided the phe-free formula is always given first[2,8].

Advantages to breast feeding are: the convenience, reduced number of bottles that need to be prepared and helps establish the mother-baby bond.

SIF fed infant

There are various options for feeding an infant with SIF + **PKU start**. The SIF may be given as the first or second feed before or after **PKU start** or **PKU start** + SIF can be mixed together in the same bottle - see section 2.6 for more detail.

Blood phe level monitoring

Blood phe levels are used to determine whether the volume of **PKU start** and BM/SIF should be adjusted. Expect to adjust the feeding plan frequently especially during the first two months of life. Initially blood phe levels should be checked twice weekly until they have stabilised to enable trends in blood phe levels to be established. The quantity of phe tolerated by infants will vary and will be guided by the blood phe levels. It is vital to investigate the possible cause for changes in the phe level before adjusting the feeding plan. It is good practice to adjust phe-free formula/natural protein intake (BM/SIF) after two consecutive blood levels, unless blood phe levels are very low or very high. See section 2.4 and 2.5 for more detail.

Progression

As the infant grows the feeding plan should be adjusted so the total feed intake always provides 150-200ml/kg/day, until weaning commences.

This 'controlled' feeding plan of **PKU start** and BM/SIF combination can continue to provide 100% of the infant's nutritional requirements until the age of 4-6 months when weaning should commence - see section 5 for more details.

N.B.

BF can continue for as long as the mother and infant wish, as long as growth and blood phe levels are satisfactory.

If BF ceases before enough phe is being taken from solid foods, then a SIF should be introduced. If the mother wishes to wean the infant from the breast then a gradual approach is recommended if possible.

Summary

If phe levels are > 1,000 µmol/L at diagnosis.

Temporarily stop the natural protein source (BM/SIF) and replace with the phe-free formula (**PKU start**).

Once phe levels are < 1000 µmol/L, reintroduce the natural protein source (BM/SIF) to tolerance.

If phe levels are 360 - 1,000 µmol/L at diagnosis.

Natural protein source does not need to be completely stopped. Introduce **PKU start** in conjunction with BF/SIF.

Natural protein intake is adjusted based on individual blood phe levels.

Initially blood phe levels should be checked twice weekly until they have stabilised.

Continue to give a combination of **PKU start** + BM/SIF to provide 100% of the infant's requirements.

This controlled feeding plan of BM/SIF and **PKU start** can continue to provide the infant's nutritional requirements until the age of 4-6 months when weaning is commenced.

The following charts show:

- Feeding regime for a BF infant + **PKU start** - Calculations are rounded to the nearest 5ml for practical reasons.
- Feeding regime for a SIF fed infant + **PKU start** - Calculations are rounded to the nearest 5ml for practical reasons.
- Check list for blood phe monitoring.
- Adjustments to the feeding plan after blood phe monitoring.
- Options for feeding the SIF fed infant + **PKU start**.

Introducing and adjusting PKU start is based on the individual, practical examples are given in this guide, however international and local practice may vary.

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

The aim is to achieve a rapid reduction in blood phe levels.

If blood phe level is > 1,000 µmol/L at diagnosis

Step 1 - Introduction of PKU start

- All breast milk should be temporarily stopped to achieve a reduction in blood phe levels below 1,000 µmol/L. Typically this should be a reduction of between 300-600 µmol/L daily.
- **PKU start** should be introduced to temporarily replace all breast feeds: it should be offered on demand to the infant.
- The infant should be weighed and as a guide **PKU start** introduced at 150ml/kg/day.
- The mother should express breast milk frequently to maintain supply whilst the infant is taking **PKU start** only.
- Blood phe is likely to decrease quickly when the prescribed volume of **PKU start** is taken. It may only take 24 to 48 hours to achieve blood phe levels below 1000 µmol/L. Therefore blood phe levels should be measured within 48 hours of introducing **PKU start**.

NB: To help maintain supply of breast milk, it is considered reasonable to introduce 1 or 2 breast feeds per day after 24 hours, even if blood phe levels are still > 1000 µmol/L. Limit each feed to a maximum of 5 minutes on the breast and offer after the prescribed PKU start is given first.

Step 2 - Reintroduction of breast milk -

Once blood phe levels reduce to < 1,000 µmol/L

The quantity of phe tolerated by infants will vary and will be guided by blood phe levels.

It is essential phe is added back into the infant's feeding regimen by giving some as BM.

If blood phe level is 360 - 1,000 µmol/L at diagnosis

Introduction of PKU start

Breast feeding does not need to be completely stopped and **PKU start** should be introduced in conjunction with breast feeding.

Breast feeding an infant with PKU is based upon the following principles which act to reduce the infant's intake of BM and therefore phe intake:

- As a starting point, the amount of **PKU start** should provide 50% of the estimated fluid requirement.
- A measured quantity of **PKU start** is given before each breast feed, this partially satisfies the infants appetite.
- The infant is then allowed to breastfeed until they are satisfied.

PKU start and breast milk should be given at the same feed to ensure the infant is provided with a balance of essential amino acids.
If the prescribed volume of PKU start is not taken, it may cause raised blood phe levels.
Initially blood phe levels should be checked twice weekly until they have stabilised to enable trends in blood phe levels to be established.
Blood phe levels are used to determine whether the volumes of PKU start/BF should be adjusted - see section 2.5 for more details.
The infant should be weighed weekly for the first 6 -8 weeks.

Example feeding plan

A 9 day old infant diagnosed with PKU, previously been exclusively breast fed. Weight 3.5kg. This calculation is based on fluid requirements of 150ml/kg/day.

Step 1 - Blood phe level is > 1,000 µmol/L

Total feed requirements:	150ml x 3.5kg = 525ml **PKU start**/day
Divide into 8 feeds/day	525ml ÷ 8 = 65ml **PKU start**/feed
New feed regimen	**65ml PKU start (x 8 feeds/day)**

Step 2 - When blood phe level is < 1,000 µmol/L

Feeding regimen	65ml **PKU start** x 8 feeds/day
The amount of **PKU start** given to the infant is halved and is given before breast feeds	½ **PKU start** = approx. 35ml/feed
New feed regimen	**35ml of PKU start + infant can then breast feed to satiety (x 8 feeds/day)**

The aim is to achieve a rapid reduction in blood phe levels.

If blood phe level is > 1,000 µmol/L at diagnosis

Step 1 - Introduction of PKU start

- All SIF should be temporarily stopped to achieve a reduction in blood phe levels below 1,000 µmol/L. Typically this should be a reduction of between 300-600 µmol/L daily.
- The infant should be weighed and as a guide **PKU start** introduced at 150ml/kg/day.
- Blood phe is likely to decrease quickly when the prescribed volume of **PKU start** is taken. It may only take 24 to 48 hours to achieve blood phe levels below 1,000 µmol/L. Therefore, blood phe levels should be measured within 48 hours of introducing **PKU start**.

Step 2 - Reintroduction of SIF -

Once blood phe levels reduced to < 1,000 µmol/L

The quantity of phe tolerated by infants will vary and will be guided by blood phe levels.

It is essential phe is added back into the infant's feeding regimen by giving a measured amount of SIF at each feed.

If blood phe level is 360 - 1,000 µmol/L at diagnosis

Introduction of PKU start

Feeding with SIF does not need to be completely stopped - **PKU start** should be introduced in conjunction with SIF.

- The infant should be weighed and total fluid intake of 150ml/kg/day maintained (as a combination of **PKU start** + SIF).
- SIF should be added into the feeding plan to provide 50mg phe/kg/day[7] and **PKU start** meets remaining nutritional requirements.
- The volume of SIF required to provide the phe requirements is divided into the same number of feeds as **PKU start**.

PKU start and SIF should be given at the same feed to ensure the infant is provided with a balance of essential amino acids.
If the prescribed volume of PKU start is not taken, it may cause raised blood phe levels.
Initially blood phe levels should be checked twice weekly until they have stabilised to enable trends in blood phe levels to be established.
Blood phe levels are used to determine whether the volumes of PKU start/SIF should be adjusted - see section 2.5 for more details.
The infant should be weighed weekly for the first 6 -8 weeks.

Example feeding plan

A 9 day old infant diagnosed with PKU, previously been totally SIF fed. Weight 3.5kg. This calculation is based on fluid requirements of 150ml/kg/day.

Step 1 - Blood phe level is > 1,000 µmol/L

Total feed requirements	150ml x 3.5kg = 525ml **PKU start**/day.
Divide into 8 feeds/day	525ml ÷ 8 = 65ml **PKU start**/feed
New feed regimen	**65ml PKU start (x 8 feeds/day)**

Step 2 - When blood phe level is < 1,000 µmol/L

Feeding regimen	65ml **PKU start** (8 x feeds/day)
phe requirement 50mg phe/kg/day[7]	3.5kg x 50mg = 175mg phe **= Approx. 230ml SIF***
Divide into 8 feeds	230 ÷ 8 = approx. 30ml SIF fed infant + feed
Infant previously required	65ml /feed (from **PKU start**)
New feed regimen (65ml/feed)	**35ml of PKU start + 30ml SIF (x 8 feeds/day)**

If the infant is still hungry after this feed, they may be offered additional **PKU start** to satiety.

*** Check phe content of the SIF being used, there is variation within formulae and this will alter the calculation. In this example the SIF provided 75mg phe per 100ml.**

2.4 Check list for 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.

Considerations for high blood phe levels:

Possible cause	Action
Excess intake of natural protein (BM/SIF)	
Incorrect understanding of the feeding plan. Inaccurate measurement or preparation of feeds.	Provide practical advice on the preparation and provision of the feeding plan.
Inadequate intake of PKU start	
Infant suffering from colic, reflux or constipation affecting tolerance and volume of **PKU start** taken. Recent vaccinations affecting tolerance and volume of **PKU start** taken. Reduced frequency or changed timings of **PKU start** (eg. stopped night feeds). Insufficient supply of **PKU start.**	Monitor weight frequently and adjust requirements accordingly. Address symptoms/seek appropriate advice to resolve barriers to tolerance. Adjust feeding regime according to tolerance. Negotiate amended feeding plan with family/ caregiver. Ensure adequate supply available.
Catabolism	
Illness or infection.	Encourage to continue with the same prescribed daily dose of both feeds but offer in smaller, more frequent volumes throughout the day. Ensure the infant has received appropriate treatment for illness/infection.
Slow rate of weight gain	Assess the overall growth trend and adjust the feeding plan as indicated. Cross-check mg/kg phe and ensure the infant is meeting minimal phe requirements. Monitor weight frequently.
Incorrect phe-free formula prescribed/taken	Confirm **PKU start** has been supplied. Show families/caregivers a can of **PKU start** to ensure they know what it looks like.
Change in routine	
Blood sample taken at a different/inconsistent time.	Agree appropriate time with families/caregivers for sample to be obtained.

In circumstances such as illness, infection or vomiting adjustments to feeding regime will be temporary.

Repeat blood phe sample within 2 days, if blood phe level is still HIGH adjust feed regimen, see section 2.5.

Considerations for low blood phe levels:

Possible cause	Action
Inadequate intake of natural protein (BM/SIF)	
Incorrect understanding of the feeding plan. Inaccurate measurement or preparation of SIF. Infant is not latching on the breast well. Infant is taking other fluids such as water replacing BM/SIF. Reduced frequency or changed timings of BM/SIF (eg. stopped night feeds/sleeping through feeds).	Provide practical advice on the preparation and provision of the feeding plan. Seek breast-feeding support. Clarify feeding plan with family/caregiver. Negotiate amended feeding plan with family/ caregiver.
Excess intake of PKU start	
PKU start given throughout the night for satiety.	Consider reducing the volume of **PKU start** overnight.
Vomiting	Encourage to continue with the same prescribed daily dose of both feeds but offer in smaller, more frequent volumes throughout the day. Ensure the infant has received appropriate treatment for illness/infection.
Anabolic	
Anabolic phase following infection.	Monitor levels. Repeat blood phe concentration and if still low, consider increasing natural protein (BM/SIF) by approximately 25-50mg phe and monitor levels carefully.
Increased rate of weight gain	Assess the overall growth trend and adjust the feeding plan as indicated. Cross-check mg/kg phe and ensure the infant is meeting minimal phe requirements. Monitor weight frequently.

In circumstances such as illness, infection or vomiting adjustments to feeding regime will be temporary.

Repeat blood phe sample within 2 days, if blood phe level is still LOW adjust feed regimen, see section 2.5.

2.5 Adjustments to the feeding plan

For breast fed infants it is important to remember once phe levels are stabilised, do not make changes to a feeding plan too frequently:

- Many factors affect phe levels; consider all causes in section 2.4
- Monitor the trend of phe levels
- In general make no more than 1 change to feeding plan in 1 week
- Consider the timing the blood sample was taken
- Consider when the last change to the feeding plan was made

For a BF infant + PKU start

If blood phe level remains HIGH after 2 consecutive samples - follow the adjustment below.

Higher than the target range:	Action
By up to 100µmol/L	Increase **PKU Start** by 10ml at each feed

By increasing the volume of **PKU start** given before each breast feed the intake of breast milk (and associated phe) will naturally reduce.

If blood phe level remains LOW after 2 consecutive samples - follow the adjustment below.

Lower than the target range:	Action
By up to 100µmol/L	Decrease **PKU Start** by 10ml at each feed

By reducing the volume of **PKU start** given before each breast feed the intake of breast milk (and associated phe) will naturally increase.

Example

Infant's weight 5kg, fluid requirement 750ml/day

Approximately 50% of fluid requirement as PKU start = 375ml.

Current total feed regimen	375ml **PKU start** + Infant can then breast feed until satiety	Current total feed regimen	375ml **PKU start** + Infant can then breast feed until satiety
Divide in to 8 feeds/day	45ml **PKU start** + Infant can then breast feed until satiety	Divide in to 8 feeds/day	45ml **PKU start** + Infant can then breast feed until satiety
Adjustment	Increase 10ml **PKU start** at each feed	Adjustment	Reduce 10ml **PKU start** at each feed
New feed regimen (x 8 feeds/day)	**55ml PKU start + Infant can then breast feed until satiety**	**New feed regimen (x 8 feeds/day)**	**35ml PKU start + Infant can then breast feed until satiety**

For SIF fed infants it is important to remember once phe levels are stabilised, do not make changes to a feeding plan too frequently:

- Many factors affect phe levels; consider all causes in section 2.4
- Monitor the trend of phe levels
- In general make no more than 1 change to feeding plan in 1 week
- Consider the timing the blood sample was taken
- Consider when the last change to the feeding plan was made

For a SIF fed infant + PKU start

If blood phe level remains HIGH after 2 consecutive samples - follow the adjustment below.

Higher than the target range:	Action
By up to 100µmol/L	Decrease phe by 25mg from total days SIF
By 100 – 200µmol/L	Decrease phe by 50mg from total days SIF

By decreasing the volume of SIF (phe) increase the volume of **PKU start** accordingly to meet fluid/nutritional requirements.

If blood phe level remains LOW after 2 consecutive samples - follow the adjustment below.

Lower than the target range:	Action
By up to 100µmol/L	Increase phe by 25 - 50mg* to the total days SIF

By increasing the volume of SIF (phe) decrease the volume of **PKU start** accordingly to meet fluid/nutritional requirements.

Example

Infant's weight 5kg, fluid requirement 750ml/day = 95ml/feed (x8/day).

phe tolerance 250mg/day = 330ml of SIF**.

Current total feed regimen	330ml SIF + 420ml **PKU start**	Current total feed regimen	330ml SIF + 420ml **PKU start**
Decrease phe intake by 25mg/day (35ml of SIF)	295ml SIF + 455ml **PKU start**	Increase phe by 25mg/day (35ml of SIF)	365ml SIF + 385ml **PKU start**
New feed regimen (x 8 feeds/day)	**35ml SIF + 60ml PKU start**	**New feed regimen (x 8 feeds/day)**	**45ml SIF + 50ml PKU start**

* **In general adjustments to increase phe intake would be in 25mg/day steps, particularly for those infants with more severe PKU. Increasing by 50mg/day may be appropriate for those with milder PKU or where blood phe levels are consistently below 30µmol/L. It is the responsibility of the managing healthcare professional to use clinical judgment when adjusting feeding plans.**

** **Check phe content of the SIF being used, there is variation within formulae and this will alter the calculation. In this example the SIF provided 75mg phe per 100ml.**

2.6 Options of feeding an infant with SIF + PKU start

There are various options of feeding an infant with SIF+ **PKU start** – choose the method which you are most confident with and that is most acceptable to the family/caregiver/infant. Whichever option you use do not change to another method as this will confuse the parent and infant.

Ensuring a minimal volume of **PKU start** is given is just as important as ensuring that all the SIF and therefore phe is taken.

SIF is given BEFORE PKU start

- Calculate the volume of both SIF and **PKU start** required.
- A measured amount of SIF is given first to ensure the entire phe source is taken.
- Followed by a measured amount of **PKU start**.
- This method will ensure the entire phe requirement is given.

SIF is given AFTER PKU start

- Calculate the volume of both SIF and **PKU start** required.
- A measured volume of **PKU start** is given first.
- Followed by a measured amount of SIF.
- This method will ensure the infant consumes the required amount of **PKU start**.
  This may be beneficial if the infant has a higher phe tolerance and therefore a higher SIF intake.

For both options:

- Both feeds are given in separate bottles.
- **PKU start** and the SIF should be given at the same feed to ensure that the infant is provided with the correct balance of all the essential amino acids.
- If the infant is still hungry after the feed offer additional **PKU start** to achieve satiety.

PKU start and SIF are mixed together in the same bottle

- Calculate the volume of both SIF and **PKU start** required.
- Mix the two feeds together and give from one feeding bottle.
- Ensure the full volume is consumed.
- If the infant is still hungry after the feed, offer additional **PKU start** to achieve satiety.

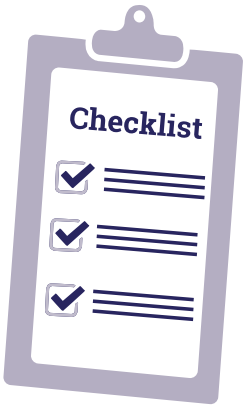
A major disadvantage of using this method is that the infant does not become accustomed to the taste of **PKU start** and therefore may find it difficult to adapt to the taste of a more concentrated second stage protein substitute which is introduced at the weaning stage.

A further issue is if any of the mixed formula is not consumed it is not possible to determine whether it is **PKU start** or SIF.

3.0 Practical points

3.1 Checklist for the healthcare professional (HCP)

- **Keep information simple and practical.**
- **Check understanding of information by the families/caregivers** and allow time for questions.
- **Discuss**
  - How blood results will be reported such as via email/text and frequency.
  - How clinics operate - number of clinic appointments and which team members they will be visiting.
- **Inform other HCPs** from the wider team of the patient management plan.
  - Such as GP; health visitor; local dietitian.
- **Organise prescription letter** to be sent to local GP.
- Set up on **home delivery** service if available.
- It is useful to give the family/caregivers **a written feeding plan** that can be used as guidance for the intake of **PKU start**.
- Ask the family/caregivers to **keep a record** of the infant's daily feed intake.
- Maintain telephone **contact**/visit the family/caregivers on a regular basis to ensure the infant is feeding well and the family/caregivers are supported.
- The breast-feeding mother will require **support** during this period by suitably qualified HCPs who specialise in breast feeding. If available provide a breast pump if necessary.
- Provide **useful contacts** and phone numbers.
- **Encourage families/caregivers** to contact dietitian/clinician for information when it comes to queries on feeding.
- Direct parents to **appropriate** websites/social media/information sites.
- Inform them of any national/local **support groups** available.



3.2 Practical points for family/caregivers for the successful feeding of PKU start and BM/SIF

Be organised – encourage parent/caregiver to be prepared e.g. bottles, amounts of powder(s) for each feed weighed out.

Routine – establish a routine of giving the chosen feed combination at the same time each day.

Complete the feeds – emphasise the importance of taking the full prescribed amount of BOTH feeds.

Teach – provide practical information regarding making up a bottle and the practicalities such as cleaning; sterilising and storing feeding equipment if mother has previously been breast feeding exclusively.

Bottle feeding – use two different coloured bottles – one for the SIF and one for **PKU start** so it is easier to distinguish between them and help reduce risk of mix-ups.

Be consistent – with the way the infant is fed and the order feeds are given amongst family members/caregivers.

Provide information for other caregivers –

- Provide the caregiver with an explanation of PKU.
- Provide the caregiver with clear written instructions of the feeding plan and the necessary feeding equipment.
- Ask the caregiver to record exactly when and how much of each feed the infant took.
- The breast fed infant with PKU can be given expressed BM on demand instead of a breast feed if with another caregiver.

4.0 Preparation guidelines

4.0

4.0 Preparation guidelines for PKU start

Only prepare one bottle at a time. Use only the scoop provided in the can of PKU start.

The standard dilution is 14.1% (14.1g of powder made up to a final volume of 100ml with water). The standard dilution is made by adding 1 level scoop (4.7g powder) of **PKU start** to 30ml of water (approx. 1 fluid oz.)

Follow the preparation guidelines carefully.

Wash hands well.

Sterilise feeding equipment according to manufacturer's instructions.

Boil fresh water and leave to cool for no more than 30 minutes to ensure it remains at a temperature of at least 70°C. Do not use artificially softened water or repeatedly boiled water.

Measure out the required amount of water into the bottle.

Using the scoop provided, add the prescribed number of scoops of **PKU start** to the water, levelling each scoop off with the back of a clean dry knife. Do not press the powder into the scoop.

Place the sterilised teat and cap on the bottle and shake well until all the powder has dissolved.

Cool to drinking temperature (approx. 37°C.) Always test the temperature before feeding by shaking a few drops onto the inside of your wrist – the feed should feel warm but not hot.

Any formula remaining in the bottle after 1 hour should be discarded.

Do not reheat **PKU start** once feeding has started.

Do not heat **PKU start** in a microwave as uneven heating may occur and could cause scalding.

Do not boil **PKU start**.

Infants should be supervised at all times when feeding.

Regular teeth cleaning is recommended.

Storage

Unopened: PKU start should be stored in a cool, dry place.

Once opened: Use within 3 weeks. Always replace container lid after use.

Use before best before date.

5.0 Progression to weaning

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During their first year, infants grow rapidly and so protein and energy needs are constantly increasing. As an infant grows, the volume of BM or SIF, plus **PKU start** needed to meet nutritional requirements becomes too high and it is impractical to expect an infant to consume that volume of liquid. The timely introduction of complementary foods during infancy is necessary for both nutritional and developmental reasons.

In all infants, the primary aim of weaning (complementary feeding) is to progressively replace an exclusively liquid diet with a mixed diet incorporating solid foods. The aim is to encourage normal feeding development by decreasing reliance on infant feeding bottles and allow appetite for foods to be established.

The same applies for an infant with PKU – progressing from an exclusively liquid diet (a combination of BM/SIF + **PKU start**), to a diet that is a combination of phe-free food + foods containing natural protein/measured amounts of phe (phe exchanges) + a more concentrated second stage protein substitute.

BM/SIF + **PKU start**

3 - 4 mealtimes per day, consisting of **phe-free foods and natural protein/phe exchanges** + **A concentrated second stage protein substitute**

When solids are introduced to an infant with PKU, feeding can become more complex for both caregivers and HCPs. Many dietary changes are instituted so that feeding meets an infant's development needs, but attaining satisfactory growth and blood phe control is essential. If this stage is managed poorly, it may negatively impact on future feeding and long term metabolic control[9].

Timing of weaning, the introduction of a concentrated second stage protein substitute, and progression onto a more textured food in infants with PKU are crucial. For the infant with PKU, weaning from 17 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-free foods, foods containing protein/phe exchanges and the concentrated second stage protein substitute whilst maintaining blood phe levels within treatment recommendations[9].

Standard weaning guidelines that are advocated for infants should be followed when possible for infants with PKU. However, there are additional components to consider in PKU.

For more detail refer to VIA (Viatflo in Association with You) - for a guide on the introduction of complementary foods and transitioning to a concentrated second stage protein substitute in infants.

6.0 References

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