PKU

A Practical Guide to Maternal PKU

Vitaflo in Association With You

Supporting education in the dietary management of rare diseases

Disclaimer

This guide should be **read in conjunction with national and local guidelines** for the dietary management of Phenylketonuria (PKU) and maternal PKU (mPKU). This guide is based on European and American guidelines for the management of PKU as well as clinical experience and best practice recommendations for the management of mPKU.

It is **for use by healthcare professionals** working with individuals with PKU.

This guide is **not for use by individuals with PKU.**

It is for general information only and must not be used as a substitute for professional medical advice.

The product information contained in this guide, although accurate at the time of publication, is subject to change.

The most current product information may be obtained by referring to product labels.

The term protein substitutes is used throughout this guide. Protein substitutes might also be known as medical formulas, PKU medical food or PKU protein supplements.

The protein substitutes (PKU express, PKU cooler, PKU air, PKU sphere), low protein foods (ProZero, Mini crackers, Vitabite, Fate) and supplementary products (DocΩmega, Tyrosine1000) discussed in this practical guide are foods for special medical purposes (FSMP) and must be used under medical supervision.

Collaborators

Vitaflo® dietitians in collaboration with:

Charlotte Ellerton RD. BSc. PgDip

Specialist Metabolic Dietitian
Charles Dent Metabolic Unit, National Hospital for Neurology and Neurosurgery, Queen Square, London.

Abbreviations

BMI – Body mass index

DHA – Docosahexaenoic acid

Hyperphe – Hyperphenylalaninemia

IBW – Ideal body weight

IEM – Inborn errors of metabolism

IQ - Intelligence quotient

LBV – Low biological value

LCP – Long-chain polyunsaturated fatty acids

LP – Low protein

MDT – Multi-disciplinary team

mPKU – Maternal PKU

PAH – Phenylalanine hydroxylase

PE – Protein equivalent

phe – Phenylalanine

PKU - Phenylketonuria

PS – Protein substitute

RNI - Reference nutrient intake

phe exchange A measured amount of a protein containing food.
A 50mg phenylalanine exchange system is used in this guideline:
1g of natural protein = 50mg phenylalanine = 1 phe exchange.

Practice on calculating phenylalanine intakes vary internationally. Local procedure should be observed and adhered to when giving individuals advice.

Foreword

Dietary management of PKU during pregnancy is complex, and demands a great deal of both the individual and healthcare professionals supporting them. Nutritional requirements differ at every stage of maternal PKU (preconception, throughout pregnancy and during lactation) and dietary recommendations must be tailored to suit each individual. Careful monitoring and specialist expertise is essential to ensure the best outcome for both mother and infant.

The Charles Dent Metabolic Unit (CDMU) at University College London Hospitals (UCLH) is one of the largest and longest established facilities for adolescents and adults with inherited metabolic diseases. The unit was founded by Charles Dent who was one of the first physicians to recognise the teratogenic effect of high maternal phe levels in 1956[1]. CDMU has now been managing mPKU for over 40 years with nearly 300 babies born to women with PKU attending the unit. All of these children are offered developmental follow up until adolescence, and outcomes are recorded in a database which was initiated in 1977. This cumulative data contributes to improved knowledge and research and helps to ensure all women with PKU and their children continue to be offered the best possible care.

mPKU dietetic care has been influenced hugely by Maggie Lilburn who was the first metabolic dietitian at the unit. She was a pioneer in this field from 1970 until her retirement in 2004, continuing in research until 2007. Her dedication to supporting individuals throughout pregnancy continues to inspire so many.

This practical guide for the dietary management of mPKU has been created as an aid for healthcare professionals and offers realistic suggestions and ideas to help with some of the challenges faced. In addition, it illustrates how Vitaflo's PKU product range can be incorporated into the diet.

Charlotte Ellerton

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1.0 Introduction to mPKU dietary management

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Women with phenylketonuria (PKU) must optimise metabolic control when planning, and during, pregnancy. High blood phenylalanine (phe) concentrations (referred to as phe levels) are teratogenic and can result in maternal PKU syndrome: microcephaly, mental retardation, cardiac defects, spontaneous abortion and low birth weight (<2500g) in the offspring[2]. Tight control is necessary due to the positive amino acid gradient across the placenta, thereby exposing the foetus to higher phe concentrations than the mother[3].

Maternal PKU syndrome can be avoided with diligent dietary management prior to conception and during pregnancy[4-6]. The European and American Guidelines recommend maintaining phe levels between 120-360µmol/L prior to conception and throughout pregnancy[7-8]. This requires individuals with PKU to restrict their dietary phe intake and consume sufficient energy, protein, and micronutrients to meet nutritional requirements.

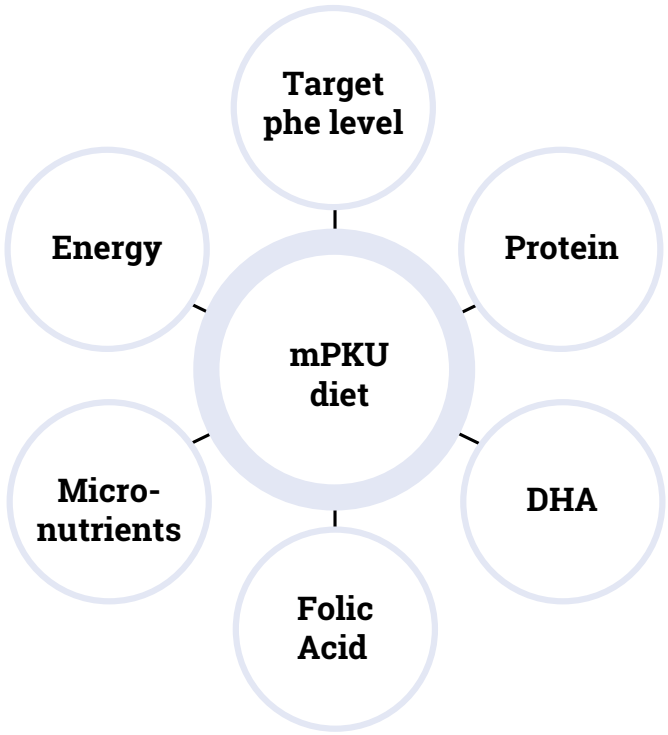
Best outcomes for children born to mothers with PKU are associated with achievement of metabolic control prior to conception[4-6]. Despite this knowledge, conception with uncontrolled phe levels (called an 'unplanned pregnancy') is common in PKU[9]. Unplanned pregnancies account for 30-60% of PKU pregnancies in large, specialist metabolic centres[10, 11]. Dietary restriction should commence immediately in the event of an unplanned pregnancy as evidence suggests that favourable birth outcomes can still occur when good metabolic control is achieved by 8–10 weeks gestation, and maintained throughout pregnancy[4].

Dietary management for a PKU pregnancy should be overseen in a specialist metabolic centre experienced in the care of maternal PKU (mPKU)[12]. Care is required from a multidisciplinary team (MDT) including a physician, dietitian, and ideally, a psychologist who are all specialised in the management of adults with inborn errors of metabolism (IEM)[7].

The aims of mPKU dietary management

1. **Reduce and maintain stable phe levels to within recommended target range**
   - The European and American PKU management guidelines recommend that phe levels are kept between 120 and 360 µmol/L preconception and throughout pregnancy[6,7].
2. **Ensure adequate nutritional intake to promote optimal growth and development of the foetus**
   - Nutritional demands change throughout preconception, pregnancy and lactation. Each stage needs careful monitoring and frequent adjustment.
   - In addition to high blood phe, suboptimal intakes of energy, protein and vitamin B_{12} have been shown to have a negative impact on outcomes[12-14].

1.1 Priorities for mPKU dietary management

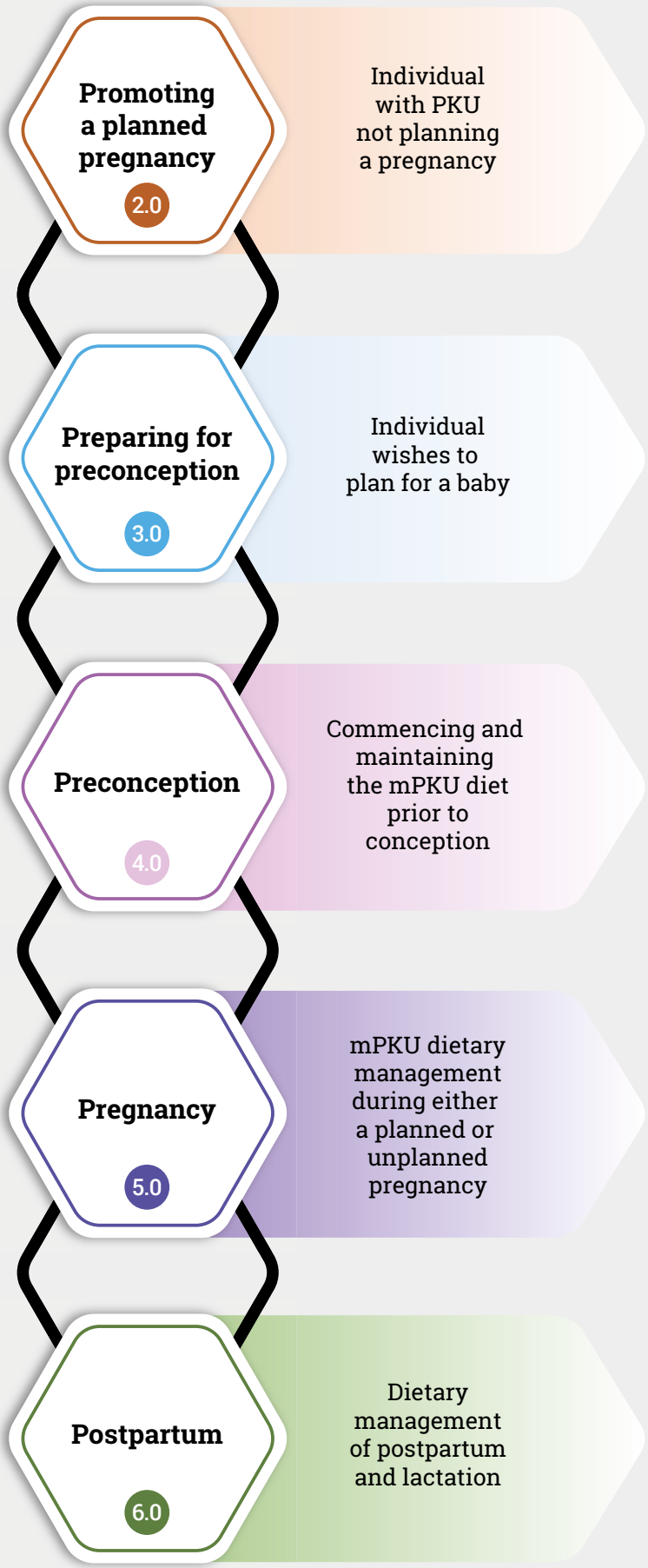


As well as managing PKU, important considerations for a healthy pregnancy include observing food hygiene (food safety), avoiding excessive vitamin A intakes and ensuring sufficient folic acid supplementation and DHA intakes.

The rest of this guide focuses on the practical management of mPKU. For further information on mPKU the following references are recommended:

- Acosta, P.B. and K. Matalon, Chapter 5: Nutrition management of individuals with inherited disorders of aromatic amino acid metabolism, in Nutrition management of individuals with inherited metabolic disorders, P.B. Acosta, Editor. 2010, Jones & Bartlett Publishers. p. 119-175.
- Acosta, P.B., et al., Intake of major nutrients by women in the Maternal Phenylketonuria (MPKU) Study and effects on plasma phenylalanine concentrations. The American journal of clinical nutrition, 2001. 73(4): p. 792-796.
- Koch, R., et al., The Maternal Phenylketonuria International Study: 1984-2002. Pediatrics, 2003. 112(6 Pt 2): p. 1523.
- Maillot, F., et al., A practical approach to maternal phenylketonuria management. Journal of inherited metabolic disease, 2007. 30(2): p. 198-201.
- Maillot, F., et al., Factors influencing outcomes in the offspring of mothers with phenylketonuria during pregnancy: the importance of variation in maternal blood phenylalanine. The American journal of clinical nutrition, 2008. 88(3): p. 700-705.
- Matalon, K.M., P.B. Acosta, and C. Azen, Role of nutrition in pregnancy with phenylketonuria and birth defects. Pediatrics, 2003. 112(6 Pt 2): p. 1534-6.
- Lee, P., et al., Maternal phenylketonuria: report from the United Kingdom Registry 1978–97. Archives of disease in childhood, 2005. 90(2): p. 143-146.

mPKU dietary management stages



2.0 Promoting a planned pregnancy

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It is important to provide continuous education to regularly reinforce the message that pregnancies in PKU should be planned[7, 9]. Knowledge and experience of managing mPKU has evolved and some women with PKU may not be aware of current recommendations, especially if they have not been attending a metabolic clinic or are 'lost to follow up'.

At regular but appropriate time points individuals with PKU and hyperphenylalaninemia (hyperphe) should be counselled about:

Effective contraception and signposting to family planning clinics as appropriate

Consequences of high phe on pregnancy outcomes

Increased risk of heart defects, microcephaly, reduced intelligence quotient (IQ), behavioural problems[2, 4-6, 16-19].

Consequences of poor adherence to protein substitutes

For the individual: Nutritional deficiencies specifically protein, vitamin B_{12}, iron and DHA[20-22].
For a foetus: Nutritional deficiencies are associated with increased risk of poor growth, microcephaly, and heart defects[13-15].

Current target phe levels for pregnancy compared to adults on diet

When treatment recommendations are adhered to, the chances of a good outcome are comparable to the general population[7].

Provide encouragement that support is available from their metabolic clinic, peers (if they would like this support) as well as their family, friends or partner (as appropriate).

Educational messages on mPKU should be embedded into care programmes at an early age[23,24]. European PKU guidelines suggest that these discussions begin at age 12. Content should be tailored to the appropriate level for the individual and their family, taking into consideration cultural or religious background and any psychological and/or intellectual disability[7].

To reinforce positive health messages of a planned pregnancy in PKU, the inclusion of partners, family or friends in the discussion should be encouraged, whilst respecting the individual's right to confidentiality[9,25]. A study of psychosocial factors in mPKU[25] found that women with PKU had more unplanned pregnancies compared to women with type 1 diabetes. They also required more support from their parents or partner to follow advice before and throughout the pregnancy[25] .

If an individual with PKU does become pregnant unexpectedly they should be encouraged to contact the metabolic centre immediately to inform them that they are pregnant or think they could be pregnant. See **unplanned PKU pregnancy section 5.4** for more information.

All individuals should also be encouraged to maintain up-to-date contact details with their metabolic centre, for themselves and their next of kin as well as ensuring that the individual is provided with up-to-date contact information for the emergency contact at the clinic.

3.0 Preparing for preconception

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In an ideal case, an individual with PKU will inform the physician or dietitian that she is ready to plan for a pregnancy. This allows for preparation of the mPKU diet, which takes time. Individuals should be provided with a realistic time frame required to achieve phe levels in the target range. Preparing for preconception involves the following stages.

Calculate

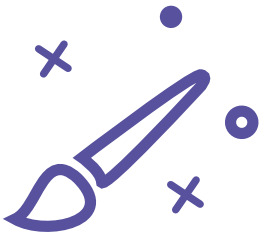
Phe, protein and other nutrientrequirements will need to be calculated prior to starting the mPKU diet for preconception. Nutritional requirements differ at every stage of mPKU. The **calculate section 7.0** illustrates requirements for preconception, during pregnancy and lactation.

Re-educate

Whether the individual is already on a phe-restricted diet, off diet or returning to care they will need education to achieve stable phe levels within the recommended target range for preconception and pregnancy.

Personalise

With any dietary intervention, tailored dietary advice will be the key to success.



Organise

Supporting the individual to plan and organise their diet will make it more manageable.

3.1 Re-educate

Individuals who are adherent to the PKU diet should be offered re-education to ensure they are following the diet precisely enough to keep their phe in the target range for preconception and pregnancy.

Intensive dietary education will be required for those who struggle with adherence or are 'off' diet.

For an unplanned pregnancy, education is required urgently.

- Several days of education will be more difficult to provide at short notice.
- Education might need to be staged, initially focusing on establishing PS and a very low phe, high calorie diet to reduce the phe level as quickly as possible.
- See the **unplanned PKU pregnancy section 5.4** for more information.

3.2 Personalise

Written information will be invaluable for individuals to take home with them.

For example phe exchange lists (each country will have their own recommendations which have not been covered in this document).

Consider different learning styles to help with understanding and retention of information.

Use visual and practical methods for education such as written information, food models, food demonstrations and supermarket trips.

Tip: Remember that the individual might not wish to tell family, friends and work colleagues that she is planning a pregnancy or is pregnant. This may impact the diet (e.g. when and where foods are weighed or PS are taken to maintain privacy).

Identification of preferred protein substitute.

Taste is very individual. Being supported and encouraged to try all the appropriate presentations of PS will help to find the option most suited to each individual. To maximise adherence, some may wish to take a combination of products to suit their taste and lifestyle.

Changes in taste preferences are common throughout pregnancy. Maintaining optimal PS and associated nutrient intakes is the priority. Altering flavour and/or preparation of PS is common during pregnancy and should be encouraged if adherence is compromised.

Tip: Encourage the individual to consider their usual routine and how the PS will be included alongside regularly spaced meals, and sufficient fluid. This includes; work, travelling, special occasions, weekends and meals with family.

There are many nutrients which need to be considered in mPKU, see section 7.0 Calculate. Some individuals may choose to take a PS with a nutrient profile which meets recommendations, limiting the number of additional supplements they may need to take.

Vitaflo's range of protein substitutes

- All PS within the Vitaflo range are designed to be interchangeable, allowing flexibility.
- All contain a comprehensive range of vitamins and minerals.
- All, except PKU express, contain DHA.

Once PS's are selected, it is essential to review the total micronutrient intake from the full PS prescription and recommend additional supplementation if required[10].

See **Vitaflo's range of protein substitutes section 8.1 and low protein foods section 8.2 for more information.**

Tip: Remember to not provide your own opinion on the taste of the PS before the individual tries it. Everyone is different and should be allowed to form their own opinion when trying the product for themselves.

Tip: Check your body language. Make sure you are open and encouraging with both your verbal and non-verbal communication.

For the management of hyperphe/mild PKU, protein restriction and/or PS may not have been required since early childhood. These individuals may have been lost to follow up and may have limited understanding of PKU and the importance of dietary restriction during pregnancy.

Individuals with hyperphe, or those who have been 'off' diet, are likely to find taking PS daily very difficult as they may not have taken a PS regularly since their childhood[7,26]. For any inidividual who has not recently taken PS routinely, it is advisable to increase the dose gradually over several days to promote tolerance and acceptance. Provide a larger sample supply of the preferred PS to take home. Allowing a longer trial of the PS should help to reduce wastage.

Identification of preferred low protein foods

It is vital for the individual to taste a variety of LP foods as these will likely provide the majority of the energy for the diet, especially during preconception and early stages of the pregnancy for individuals with a low phe tolerance. Variety is important to avoid taste fatigue and so that individuals will not be as tempted by restricted foods[27]. Providing a practical session where the individual can try and cook with the foods for themselves is invaluable.

Tip: Batch-cook LP meals and label with the date, name of dish and number of exchanges per portion before freezing. Remind patients to observe food hygiene precautions.

The individual will need enough LP foods whilst limiting wastage. Help the individual to work through their usual week and when they would likely use staples such as bread, rice, pizza bases or pasta as a base for their meals.

Recipes ideas

It is useful for the individual to leave the dietary education session with at least 3 to 4 recipes for main meals which they would feel able to cook on their own at home. Providing the necessary LP products for these recipes as samples will allow them to practice at home to build their confidence with LP cooking.

A range of delicious recipes are available from the Vitaflo Vitafriends PKU website: **www.vitafriendspku.com.** Some of these recipes require little or no specifically manufactured LP products and so are especially useful when access to products is restricted.

3.3 Organise

A continuous supply of preferred PS and LP foods needs to be set up and maintained.

Women who have been 'lost to follow up', have been 'off' diet or have hyperphe may have never independently organised their LP diet before as their parents/guardians managed it for them.

Encourage the individual to set reminders to regularly review and rotate their stock at home to ensure adequate supply within the expiry date.

Menu planning

Consolidating the education session with menu planning helps to bring all the information together. The individual can visualise what the mPKU diet will look like for them on a day to day basis. It also allows the dietitian or dietetic assistant to check their understanding of phe exchange counting and energy intake.

Essential equipment: Digital weighing scales - lightweight scales are easier to transport.

Tip: Individuals should be encouraged to carry their scales with them to accurately calculate phe exchanges when eating away from home.

Tips on eating out for the mPKU diet	Tips on holidays for the mPKU diet
Research menu options and nutritional breakdown on restaurant/café/food outlet website.	Self-catering accommodation provides flexibility.
Contact the restaurant/café/food outlet in advance to see if they will use LP pasta, rice, pizza bases in their dishes.	If in catered accommodation, ask for information on the menu, if adaptations can be made, and take advantage of the salad buffet.
Ask for possible meal adaptations such as removing phe exchange containing ingredients or changing dressings.	Encourage the individual to travel with a small supply of PS and LP foods in hand luggage.
Take digital scales to weigh out phe exchanges accurately.	Provide a customs letter for hand luggage contents.
Gluten free alternatives to bread, pasta or pizza are usually lower in protein. Some food outlets may be able to adapt gluten free options to provide a lower protein choice which could be counted into the individual's phe allowance.	Consider a short-term change to a powdered, light weight and easy to pack product such as PKU express.
	Recipes for common brands varies between countries. Encourage individuals to check the labels for aspartame, protein content and ingredients while abroad as the product's recipe may be unsuitable in the country they are visiting.

4.0 Preconception

4.0 Preconception

The best outcomes for children born to mothers with PKU are reported when phe level is controlled prior to conception and throughout pregnancy, referred to as a 'planned' pregnancy[2, 4, 12, 28].

Previous sections of this guide have covered best practice to promote a planned pregnancy as well as preparation for preconception. Once these preparations have been completed, and the individual has received sufficient supply of PS and LP foods, they will be able to commence the mPKU diet and the preconception stage.

Encourage continued use of contraception until phe levels have been stable in the target range for at least 2 weeks[7] and the individual is confident with the diet.

Commence mPKU diet.

Bloodspot monitoring - minimum once weekly[7, 29].

Some centres recommend twice weekly[9] for more frequent feedback and to encourage the diet to be followed consistently throughout the week.

Bloodspot results should be reported as quickly as possible[7].

Once to twice weekly telephone/email review[10].

Adjust the diet if phe level has not reduced into target range by 7-10 days after starting the diet (see Troubleshooting section 5.5).

Reassure that a rise in phe is expected around menstruation. No dietary change is required to anticipate this, as levels should reduce back into range afterwards (provided dietary intake remains stable).

Routine metabolic outpatient clinic appointment every 6-12 months [7, 29].

Full plasma amino acid and micronutrient profile.

3-day food diary or 24 hour dietary recall to assess phe exchanges, energy and PS intake.

Review supply of PS and LP foods and amend if additional LP foods are required for energy or variety.

Note: Referral to a fertility service should be considered for women who have maintained their phe level within the target range, and have not conceived after 6 months of stopping contraception [7, 10].

5.0 Pregnancy

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Dietary management for an unplanned pregnancy requires some additional considerations which are covered in more detail in the **unplanned PKU pregnancy section 5.4**.

Whether a planned or an unplanned pregnancy, the individuals should be encouraged to contact the metabolic clinic as soon as possible to report a positive pregnancy result. Additional monitoring for the mPKU diet during pregnancy is required.

Bloodspot monitoring - minimum twice weekly[7].

Some centres request three times weekly bloodspots[10].

Report result as soon as possible[7], ideally within 2 days of the bloodspot being taken[10].

Consider adjusting the diet if phe is within 50µmol/L of the limit of target range.
See **Troubleshooting section 5.5** for more information.

Telephone/email review twice[7,29] to three times weekly[10].

Encourage the individual to contact dietitian between reviews with questions or symptoms affecting appetite or adherence to PS or the low protein diet.

Outpatient clinic review once per trimester[7, 10, 29].

Full plasma amino acid and micronutrient profile if indicated.

Nutrition visit in clinic including weight, 3-day food diary or 24-hour recall to assess phe, energy and PS intake.

Review supply of PS and LP food and adjust as indicated.

5.1 Maternity care

A pregnancy is usually confirmed by a home pregnancy test or testing by a family physician or obstetrician. The individual should inform their family physician of pregnancy in order to initiate referral to conventional maternity services.

Planned pregnancy with good metabolic control	Unplanned pregnancy or poor metabolic control during pregnancy
The European guidelines state that pregnancy in PKU is considered high-risk as it can be difficult to maintain phe within the tight target range, therefore women are referred to an Obstetrician[7].	For women with PKU who have an 'unplanned' pregnancy, an additional early dating scan may be recommended to date the pregnancy.
Pregnancy care for a woman with PKU with good metabolic control is no different to that of a woman without PKU[10, 30].	Screening for organ development at 18-22 week ultrasound scan is recommended[7] if metabolic control or adherence to dietary advice is poor, even if phe was in target range at time of conception.
Providing there are no obstetric concerns, no additional birthing plans or procedures women, with PKU can deliver locally[31].	Monitoring, care and delivery plans should be guided by a specialist materno-foetal medicine unit if concerns are raised during obstetric monitoring[31].

To provide clear communication, a monthly summary letter providing all the bloodspot results and dietary advice given can be addressed to the individual and all professionals involved in their mPKU care. See **section 10.2 Example monthly summary letter** for more information.

Priorities for dietary management

First trimester 0 to 12 weeks of pregnancy

Good metabolic control in the early weeks of pregnancy is important for preventing foetal cardiac defects and protecting IQ[2, 6]

Dietary phe tolerance is low and so the diet will be the most restrictive during this time. Phe levels can be very sensitive.

Second and third trimester 13-40 weeks of pregnancy

After approximately 16 weeks' gestation phe tolerance can increase rapidly as foetal growth accelerates[10].

As the baby grows it is necessary to increase phe exchanges significantly to prevent the phe level reducing below the lower limit of the target range[10]. Persistent low phe levels, below 100µmol/L, have been associated with growth retardation[32].

Assess dietary adequacy. Increase dietary phe prescription by at least 1-2 phe exchanges at a time when phe reduces to below 150µmol/L on 1-2 consecutive bloodspots. At times, this increase may need to be more rapid, in line with growth, to prevent phe dropping below 120µmol/L. For more information see **Troubleshooting section 5.5**.

Some women find increasing phe exchanges in their diet difficult and will require encouragement and inspiration. When dietary phe is 15 phe exchanges or above, protein foods can be included. Portions of 3-6 phe exchanges are useful[33]. For recipes which provide directions on how to add or remove phe exchanges to suit the individual ask your Vitaflo representative for the Live Life Well recipe pack.

Pregnancy associated nausea and vomiting, viruses, infections, and changes in appetite can have a detrimental effect on metabolic control[28] due to inability to meet energy and/or protein requirements. This can lead to catabolism and stimulate release of endogenous protein stores which contribute to elevated phe levels[13]. Preventing catabolism during this time is essential, see **sections 5.3 Nausea and vomiting and 5.5 Troubleshooting** for more information.

Providing enough energy, protein and micro-nutrients is essential to support growth and development[14, 28].

Aim to maintain stable levels within the target range. Variation in phe level has been shown to be negatively associated with birth weight[28].

Gestational stages of development

10 weeks

The heart is fully formed by 10 weeks and has already started to beat.

12 weeks

By 12 weeks the organs of the foetus including the brain are formed. The foetus will be about the size of a lemon.

Most women will not know they are pregnant until they miss their first or second menses (period) which relates to the 4th or 8th week of pregnancy.

20 weeks

By 20 weeks the foetus is about the same length as a carrot.

30 weeks

By 30 weeks the foetus is about the size of a pineapple.

Pregnancy associated nausea and vomiting usually lessens after 12-16 weeks gestation[34], however, symptoms of indigestion, heart-burn, dizziness and constipation are more common after week 20.

5.3 Nausea and vomiting

Nausea and vomiting is common, affecting up to 85% of all pregnancies[34]. Symptoms typically subside between 12-16 weeks but for up to 15% of women the symptoms will persist beyond 16 weeks[34]. These symptoms can affect appetite[34] but women also often describe feelings of isolation, fatigue, helplessness, depression, anxiety, frustration, difficulty in coping and irritability[34].

For PKU, this can mean poor adherence to PS, difficulty following a strict diet and/or inability to consume sufficient calories to prevent catabolism[28]. This can lead to increasing phe levels as well as insufficient protein and micronutrient intakes. Symptoms of nausea and vomiting in mPKU need to be identified promptly and treated aggressively[7].

Advice from the family physician or obstetrician should be sought. Acid-reducing and anti-emetic medications are often discussed early and recommended at a low threshold[7]. Anti-emetic medications are more effective if used earlier[35] and effective use helps to regain metabolic control if it is compromised by nausea, vomiting and associated poor appetite.

In extreme cases, enteral tube feeding can be considered. Positive outcomes have been reported in 2 cases where gastrostomy tube feeding was used for management of mPKU[36]. Gastrostomy tube placements during pregnancy are complicated by the potential risk of uterine and foetal injury[37]. Each case should be reviewed individually to assess benefit versus risk of each intervention.

If the individual vomits after taking their protein substitute they must take another dose as soon as possible to minimise any rise in phe level.

General tips for nausea in pregnancy[35]:	Practical tips to help adherence to protein substitutes (PS):
Small, frequent meals and drinks Eating a small meal or snack every 2-3 hours may be helpful. Remember, if portion sizes are smaller, extra calories need to be included elsewhere in the diet e.g. food fortification, high calorie drinks, or snacks.	**Avoid the smell** Use ready to drink resealable PS such as PKU cooler or PKU air. For PKU express and PKU sphere* use a beaker with a fitted straw to make up the PS or cover the glass with plastic wrap and pierce with a straw.
Cold meals This is particularly useful if smells are triggering nausea. LP sandwiches, LP pasta or LP rice salad can be helpful.	**Take the PS very cold** Make PKU express or PKU sphere up with ice-cold water or keep PKU cooler or PKU air in the fridge. Mini cool packs can be useful to keep the PS cool during the day whilst away from home.
Eat plain carbohydrate-rich foods 20 minutes before getting up in the morning Snacking on plain, carbohydrate-rich foods such as Mini crackers or plain LP toast before getting up might help to reduce nausea.	**Try taking small amounts more frequently** Split into smaller doses more regularly through the day. This might be particularly useful if there is a certain time of the day when nausea is worse. Smaller pack sizes are available for certain PS so smaller doses can be carried and taken more conveniently.
Try ginger Many women say ginger can reduce nausea. In PKU suitable sources include fresh or crystallised ginger, ginger tea, non-alcoholic ginger beer (make sure the label is checked for protein or aspartame).	**Try a different dilution** Some women find a more dilute PS easier to take and so, additional water or permitted squash/drink could be added to PKU express. PKU express can be made with less fluid and taken as a 'shot', or 'mini drink' followed by a drink of water, or permitted juice/squash.
Enough rest The need for sleep increases during early pregnancy, and being tired may make nausea and vomiting during pregnancy worse. Encourage the individual to let their family or friends know how they can help[38].	**Try a different flavour** Sometimes a change in flavour or PS is needed.
	Try a different preparation PS are available as ready to drink liquids, powders (in sachets or tins), and as tablets.

*PKU sphere contains phe and so caution should be used when incorporating into the mPKU diet. Changes should be made gradually and closely monitored. It is suggested that dietary phe intake should be adjusted to account for the phe content from GMP-based PS.

5.4 Unplanned PKU pregnancy

Dietary restriction should commence immediately in unplanned pregnancies and women should be offered to be seen within 24 hours of informing the metabolic unit of the pregnancy[24]. This appointment should include a full clinician's assessment and dietary education.

Where an individual is unable to attend an appointment, advice can be provided over the phone and/or email. You could use the **Information collection checklist** and **Dietary advice checklist** on the following pages to support this conversation.

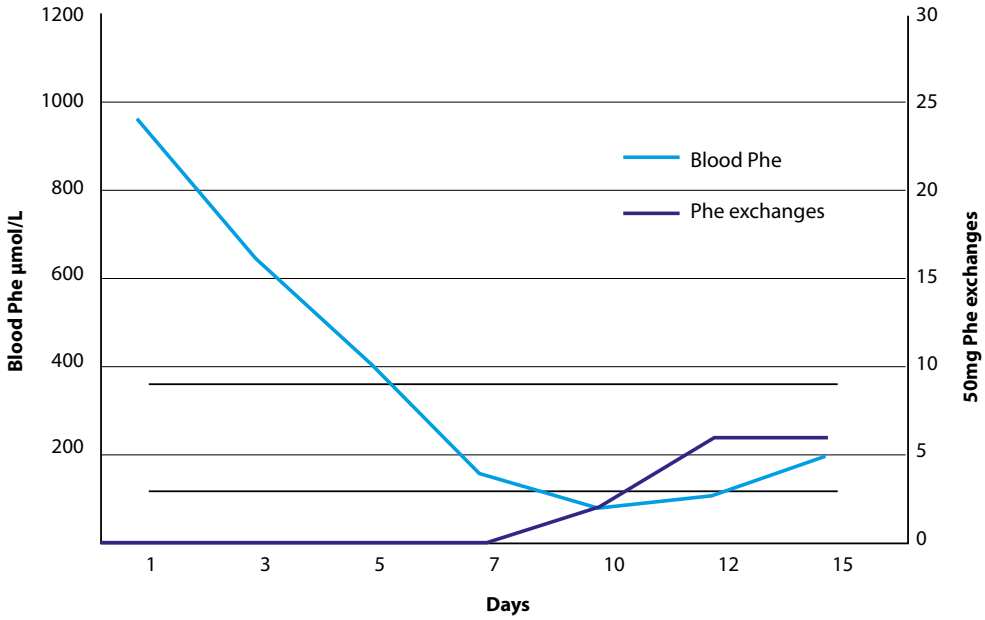
In 'unplanned' pregnancies, evidence suggests that more favourable birth outcomes are reported if the phe level is between 120 and 360µmol/L at the earliest possible point[12, 28].

Studies show that the later a woman with PKU gains control of her phe levels during pregnancy, the more likely it is that the child will have[2, 12, 28, 37, 38]:

- microcephaly
- lower intelligence/IQ
- cardiac defects
- behavioural problems such as aggression and poor attention

Achieving metabolic control by 8–10 weeks gestation is critical[4]. If metabolic control is not achieved by 10 weeks' gestation the Metabolic Consultant may counsel the individual to discuss options around continuing the pregnancy[10].

When the individual is established on a low phe diet with sufficient PS, the phe levels should reduce quickly into the target range (typically within 7 days). To prevent the phe level dropping below the lower limit of the target range phe exchanges will need to be reintroduced. For individuals with milder PKU or hyperphe, the reintroduction of dietary phe might need to be rapid, possibly requiring an increase of up to 4-6 phe exchanges at a time.



Information collection check list

When the individual makes contact to report an unplanned pregnancy, collecting the information listed below can help to establish the best course of action until the individual can attend the metabolic unit.

- [ ] **Date of last period**
- [ ] **Information about any previous pregnancies**
- [ ] **Number of phe exchanges if on diet or when last on diet**
- [ ] **Stock of in-date protein substitute (PS) at home**
  - Exact brand and flavour
  - How much protein equivalent it provides
  - If it contains micronutrients and DHA
- [ ] **Last known phe level (might be several years ago)**
- [ ] **Centre previously providing care, if known**
- [ ] **24-hour dietary recall to establish:**
  - Dietary pattern
  - Phe intake
  - Energy intake
  - Knowledge of counting phe exchanges
- [ ] **Ability to take a bloodspot on their own, supply of bloodspot cards and lancets at home**
- [ ] **Partner or family who might be able to support with the diet/bloodspots**
- [ ] **Contact details including :**
  - Current home address and phone number
  - Family physician
  - Next of kin
  - Address suitable for delivery of samples and letters

Dietary advice check list

- [ ] **Establish protein substitute (PS) to provide at least 70-80g protein equivalent (PE)**
  - This is critical to reduce phe level rapidly and to ensure to meet protein and micronutrient requirements (see **section 7.0 Calculate**)
  - Ideally, the individual will have sufficient supply at home
  - If the individual has no access to PS and is unable to attend the metabolic centre, urgent samples should be ordered as a priority, ideally with next or same day delivery
- [ ] **Minimise dietary sources of phe**
  - Try to guide the individual through each meal to suggest low protein (LP) alternatives to their current meals
  - Ideally, the individual will have sufficient supply of LP products at home
  - If the patient has no access to LP foods, samples can be ordered from manufacturers or collected by the individual/posted from the metabolic unit
- [ ] **Maintain sufficient energy**
  - Adequate energy intake is vital to promote anabolism which will help to reduce phe level
  - A phe-restricted diet, without sufficient LP foods, will be low in calories
  - Encourage fortification of meals, snacks and drinks with fats, oils and sugars
  - Supplementation with glucose polymer or high fat supplements may be indicated
- [ ] **Establish bloodspot monitoring**
  - Twice to three times weekly phe bloodspots should be established as quickly as possible
  - Remind the individual to pay the appropriate postage when they post their bloodspot
  - If the individual has no access to bloodspot cards and lancets, provide a supply as quickly as possible
  - If the individual feels unable to take their own bloodspot due to needle-phobia, encourage them to seek help from a relative/friend/partner who could help them. There are demonstration videos on YouTube which you might feel are suitable to signpost to the individual.

5.5 Troubleshooting

With frequent monitoring, it will be possible to see the phe level trending upward or downward within the target range. Always check for causes of high or low blood phe level before making changes to dietary recommendations. It is good practice to adjust PS or phe exchanges after assessing two consecutive bloodspot results. If blood phe is very high, very low or the patient has contacted to report onset of illness, immediate adjustments to phe exchanges and PS should be considered.

Table 1: High phe levels – Potential causes and corrective actions

Potential cause	Action
Inadequate protein substitute (PS) consumed	Explore reasons why intake has decreased (e.g. illness). Check correct PS has been issued. Reiterate importance of PS (as well as phe control) to support growth. If the volume is too large consider using a powdered version (such PKU express) which can be concentrated, or split into more frequent, smaller doses. Offer a change in flavour or preparation of PS. Encourage discussion of underlying anxieties.
Excess phe intake	Explore reasons why phe has increased (e.g. new foods or drinks which they may not realise contain phe). Review portion sizes. Food diaries can be a useful way to identify additional phe and counting accuracy. Check low protein food has not been confused with gluten free. Encourage menu planning, aiming to spread phe exchanges out during the day. Offer further dietary education on phe counting.
Catabolism Illness or infection. If nauseous see **nausea and vomiting section**.	Reduce phe exchanges. Increase energy intake (see below). Consider increasing PS if not already maximised.
Poor energy intake or weight loss It can be difficult to meet energy requirements during preconception and early stages of pregnancy, especially when phe allowance is low	Utilise LP foods to increase variety and to prevent hunger or temptations. Increase energy in the diet by the addition of fats, oils, or sugar. Glucose polymers or high fat supplements may be required. Consider using ProZero or other permitted, high calorie fluids to make up PKU express or PKU sphere to increase energy content. Ask your Vitaflo representative for 'My PKU express recipes' and 'PKU sphere shake' recipes where appropriate. Consider increasing PS if not already maximised.
Poor adherence	Encourage discussion. Provide practical tips to address cause e.g. eating out or away from home, at work or on holidays. Involve other professionals to provide additional support and encouragement.

Table 2: Low phe levels – Potential causes and corrective actions

Potential cause	Action
Excess protein substitute (PS) consumed	Explore reasons why the intake has increased. Check correct LP food and PS has been issued. If in the third trimester and recommended > 30 phe exchanges, consider reducing PS.
Insufficient dietary phe intake	Check all recommended phe exchanges are being consumed. Suggest replacing low protein foods with protein containing sources e.g. bread, pasta, rice, couscous. Recommend foods higher in protein as tolerated, e.g. beans, pulses, dairy products. Assess if the patient is overestimating phe exchanges or basing calculation on uncooked or frozen foods.
Excess weight gain	Encourage healthy weight gain. Food diaries can be a very useful way to assess for excessive energy intake. Consider a lower sugar and calorie PS if appropriate. Encourage use of phe exchanges on staple foods or high protein foods to reduce overall calorie intake.
Rapid foetal growth From 16 weeks of pregnancy phe tolerance/ requirement increases alongside foetal growth.	At around 18-25 weeks phe exchanges may need to be increased significantly in a short period as growth accelerates. Increase dietary phe prescription by at least 1-2 exchanges at a time when phe reduces to below 150µmol/L on 2 consecutive bloodspots. This increase may need to be more rapid in line with rapidly reducing phe levels to prevent phe dropping below 120 µmol/L. Recommend higher protein foods in 3-6 phe exchange portions once dietary phe reaches 15 phe exchanges

Table 3: phe exchanges not increasing after 16-20 weeks' gestation – Potential causes and corrective actions

Potential cause	Action
Poor adherence As phe tolerance increases the individual may find she is able to consume more phe without adverse effects on phe control. It is important for both the dietitian and individual to know the number of phe exchanges being consumed so that accurate changes can be made if needed.	Ask the individual to complete a food diary with their actual intake. For example, the individual reports consuming 8 phe exchanges, food diary reveals they are consuming 12 phe exchanges, phe level is stable in target range, then encourage the individual to weigh and measure out 12 phe exchanges.
Inadequate protein substitute consumed	Explore reasons why intake has decreased (e.g. illness, high volume). Check correct PS have been issued. Reiterate importance of PS to support growth as well as phe control. If the volume is too large consider using a powdered version (such PKU express or PKU sphere*) which can be concentrated, or split into more frequent, smaller doses. Encourage discussion of underlying anxieties.
Poor foetal growth If the baby stops growing phe tolerance reduces and phe level can rise.	Ensure to inform all healthcare professionals involved in their care of high phe levels, especially if there are already concerns of poor foetal growth.
Poor energy intake or weight loss	Utilise LP foods to increase energy intake. Ask if they have taste preference or cravings for certain foods and suggest new recipes. Increase energy in the diet by the addition of fats, oils, or sugar. Glucose polymers or high fat supplements may be required. Consider using ProZero or other permitted, high calorie fluids to make up PKU express or PKU sphere to increase energy content. Ask your Vitaflo representative for 'My PKU Express Recipes'. Consider increasing PS if not already maximised.

*PKU sphere contains phe and so caution should be used when incorporating into the mPKU diet. Changes should be made gradually and closely monitored. It is suggested that dietary phe intake should be adjusted to account for the phe content from GMP-based PS.

In the experience of CDMU, individuals who have an uncontrolled phe during pregnancy should be:

- highlighted to the metabolic physician
- offered a metabolic clinic appointment, further dietary education and/or a home visit to help identify factors affecting metabolic control
- offered a hospital admission to supervise the low protein diet, intake of protein substitutes and bloodspot monitoring as well as providing further advice and support from the dietitians, dietetic assistant, metabolic physician, or psychologist as required.

If phe levels remain high after 2 weeks of interventions:

- a MDT meeting should be called to discuss next actions including referral to appropriate services if there are safeguarding concerns.
- the outcome of the MDT meeting should be discussed with the individual.

In the CDMU's experience safeguarding services have; reinforced the health messages from the metabolic team, offered additional support such as childcare, provided some financial support with attending appointments, and supported with posting of bloodspots. The process of involving safeguarding services is lengthy and the MDT decision to refer is not taken lightly due to concern of breaking trust or rapport between the metabolic team and the individual.

6.0 Postpartum

6.0 Postpartum

For the infant	For the mother
Record weight, length (if available) and head circumference at birth. Newborn screening as per national/local protocol. An echocardiogram is recommended for all infants where metabolic control was poor at conception or during the pregnancy[7]. Follow up care with psychometric assessments by a clinical psychologist at 1 year, 4 years and 8 years is beneficial[10].	Recommence contraception after birth. It has been observed that women with PKU who did not plan their pregnancy may be less likely to plan subsequent pregnancies[28]. Offer a metabolic outpatient clinic review 4-8 weeks post partum [10, 29], and routinely thereafter 6-12 monthly[7, 29]. Usual follow up from maternity services[7] e.g. health visitor and family physician.

Exclusive breastfeeding for a baby's first 6 months of life, where appropriate, should be encouraged in line with WHO recommendations[41].

Mothers with PKU should be encouraged to breastfeed irrespective of their phe level postpartum[33, 42]. Infants born to women with PKU who do not inherit PKU are able to metabolise phe contained in their mother's breastmilk without difficulty[7,30, 42].

Babies who do inherit PKU can still be breastfed by their mother, in conjunction with phe-free infant formula, with close monitoring[7].

Benefits of breastfeeding for the infant	Benefits of breastfeeding for the mother
• Providing ideal nutrition for healthy growth and development[41]. • Helping to build up a strong immune system[41]. • Encouraging a strong bond between mother and infant[41].	• Faster return to pre-pregnancy weight[41]. • Protection against certain diseases[41].

Adherence to the PKU diet can be particularly challenging after the baby is born. The new baby will likely require much of the individual's time and attention.

Continuing the PKU diet

Benefits for adults aiming for phe levels within the target range include:

- improved sustained attention[43]
- improved mood[43]
- improved reaction times[44]
- improved cognitive ability[45]
- improved executive function[45]

Diet should be adjusted to achieve target blood phe values for adults with PKU.

Tip: Preparing and freezing LP meals as well as stocking up on LP foods before the baby arrives will help.

Tip: Recruit the partner or family members to support the individual with preparing and organising the diet.

It is important to ensure their diet is adequate in calories, protein and micronutrients to support lactation. See **Calculate section 7.0** for more information.

Tip: Encourage the individual to contact the centre if she would like further support.

Remember to update any ongoing supply of PS and LP foods.

7.0 Calculate

7.0 Calculate

7.1 Phenylalanine

Phe is an essential amino acid so a small, measured amount needs to be provided from the diet. Phe requirement/tolerance varies between individuals depending on their PAH enzyme activity level, and can also be affected by[13,46]:

- Adequacy of PS intake
- State of health (i.e. presence of nausea, vomiting or infection)
- Stage of pregnancy
- Weight changes
- Energy intake

The initial phe prescription provides a starting point for preconception. The aim is to maintain stable levels in the middle of the target range prior to conception. After initiating the mPKU diet for preconception the phe level should reduce into the target range by the 7th to 10th day[33]. If not, then further dietary manipulation may be required, see **Troubleshooting section 5.5**.

Individuals with low phe tolerance

Dietary protein intake is severely restricted to control phe intake for individuals with a low phe tolerance (previously described as 'classical' PKU).

On diet

A food diary might be useful to correlate current phe levels and phe intake and make the necessary adjustments to achieve target phe levels.

In individuals who are on diet and maintaining phe levels 500-600µmol/L, reducing their phe prescription by half should be sufficient to bring the levels into the middle of the target range 120-360µmol/L.

Off diet

If an 'off diet' blood phe level is available, it is useful for calculating the initial prescription for phe during preconception:

Table 4: suggested Phenylalanine prescription to achieve phe value 100-300µmol/L in PKU. Adapted from Maillot et al[10] and updated with CDMU experience.

Blood phe concentration – off diet	Number of phe exchanges to commence preconception diet
>2000µmol/L	4
1600-2000µmol/L	6
1200-1600µmol/L	8
1000-1200µmol/L	10 - 12
600-1000µmol/L	12 - 14

Individuals with higher phe tolerance

Individuals with high phe tolerance (previously referred to as mild PKU) or hyperphe will require less dietary restriction, however they require the same level of monitoring and support to ensure phe levels remain in target range and their dietary intake is nutritionally adequate.

Prior to making dietary changes, a series of fasting bloodspots (at least 3 to find an average) will be needed to estimate their phe tolerance. This average phe level can allow an estimation of their phe prescription using table 5.

Table 5: suggested Phenylalanine prescription to achieve phe value 100-300µmol/L in hyperphe *from CDMU experience.

Blood phe concentration – off diet	Dietary recommendations
450-600µmol/L	30-40g dietary protein and additional protein substitutes
300-450µmol/L	Usual diet and additional protein substitutes
<300µmol/L	Usual diet and monitor blood phe levels

European guidelines state that untreated phe levels of less than 360µmol/L do not require dietary management[7] but would benefit from monitoring of phe levels during pregnancy and outcome of the child to support future research.

Waisbren et al. reported the offspring of women with mild hyperphe had cognitive and behavioural outcomes similar to control children[5]. Levy et al. found that women with untreated phe <400µmol/L did not require dietary restriction during pregnancy[47]. Later Levy et al. concluded that untreated phe <600 µmol/L in an untreated individual was not overtly teratogenic and does not require dietary therapy[48].

7.2 Protein

Due to the restriction of dietary protein required to achieve target phe levels in mPKU, it is vital to ensure optimal protein substitute (PS) is prescribed to meet protein requirements[13, 15].

The role of PS in dietary management of mPKU includes:

- Lowering the blood phe concentration[7, 13]
- Providing a main source of protein and micronutrients which are essential for growth and cardiac development of the foetus[13-15, 49].

To promote protein utilisation and anabolism PS should be consumed:

- At regular intervals in 3-4 doses spread throughout the day[50]
- With LP sources of energy[51]

Protein requirements increase in pregnancy and this usually complements an increase in phe tolerance. This increase in phe tolerance is therefore met by dietary protein rather than increasing PS. As phe tolerance increases the individual can be encouraged to include higher protein options as tolerated[10, 33].

There are several sources which advise on calculation of protein requirements in mPKU which are summarised in table 6.

Table 6: Calculation of protein requirements for mPKU

Reference	Calculation for Daily Protein Requirements (g/Day)				
	Non-pregnant adult	**First trimester**	**Second trimester**	**Third trimester**	**Lactation (up to 6 months)**
European PKU guidelines[7]	0.83g protein/kg minus intake of natural protein + 40%	>70	>70	>70	Non-pregnant adult calculation + 15g
American PKU guidelines[29]	120-140% RDA for age	>70	>70	>70	>70
Acosta[46]	70	70	85	100	Not specified
Martiz[33]	n/a	59 (RNI for pregnancy (51g) + 15%)	Not specified	Not specified	Not specified

Table 7: Calculation of protein requirements for pregnancy for the general population

Reference	Calculation for Daily Protein Requirements (g/Day)				
	Non-pregnant adult	**First trimester**	**Second trimester**	**Third trimester**	**Lactation (up to 6 months)**
FAO/WHO/ UNU 2007[52]	0.83g protein/kg	+1	+10	+31	+19

This calculation assumes that all protein consumed is of good quality. In PKU, a factor of 20% to compensate for the 'digestible indispensable amino acid score' is added[7, 52]. European guidelines recommend an additional 20% is added to optimise their impact on phe control[7].

Practical tips:

- Round dose of PS up to the nearest 10g
- Ideal body weight IBW used for calculations if BMI > 25kg/m²

Example scenario:

A 27 year old woman contacts the unit wishing to plan a pregnancy

Weight: 65kg Height: 1.68m BMI: 23kg/m²

Example calculation: FAO/WHO/UNU 2007 recommend safe level of protein intake for adults is 0.83g protein per kg body weight per day. This calculation assumes that all protein consumed is of good quality. In PKU, a factor of 20% for the indigestibility of amino acid PS is added.

Weight (kg)	x	**0.83**	x	**20%**	=	**PE required from PS and phe exchanges for adults with PKU.**
65kg	x	**0.83**	x	**1.2**	=	**65g**

Off diet phe level: 1600µmol/L

Prescription of phe (using table 4): 6 phe exchanges

65g	-	**6g**	=	**59g**
Total protein requirements for Miss Brown		Phe exchange requirement		PE required from PS Rounded up to the nearest 10g.

This individual would therefore need to be prescribed **60g PE** from a PS of her choice.

Twin pregnancy

No research has been published regarding twin pregnancies in individuals with PKU. Maillot et al. stated, in their experience, keeping up with protein requirement and phe requirement during the second and third trimester of a twin PKU pregnancy is challenging[10].

FAO/WHO/UNU indicate an additional 50g protein and 1000kcal after 20 weeks gestation in non-PKU twin pregnancies may be beneficial[52].

7.3 Energy

Providing enough energy prevents catabolism and therefore helps to maintain a more stable phe level. Inadequate energy intake is linked with poor maternal weight gain, lower birth weight and poorer phe control[13, 15].

There are several reports which estimate the additional energy demands of pregnancy and lactation for the general population. No studies currently have determined energy requirements in pregnancy or lactation specifically for PKU.

Table 8: Energy requirement recommendations for pregnancy and lactation in the general population

Reference	Estimated Additional Energy Required (kcal/Day)			
	First trimester	**Second trimester**	**Third trimester**	**Lactation (up to 6 months)**
EFSA 2013[53]	+70	+260	+500	+500
SACN 2011[54]	nil	nil	+191	+330
IOM 2005[55] **for girls 14-18 and women 19-50 years**	nil	+340	+454	+330
FAO/WHO/UNU 2001[56]	+85	+360	+475	+505

As rates of overweight and obesity in the general and PKU population are rising[57], it is important to also avoid excess weight gain during pregnancy. It is ideal for women to begin pregnancy at a healthy weight (BMI 18.5-24.9 kg/m²). Women who are underweight or overweight at the beginning of pregnancy are at risk of poor maternal and foetal outcomes. Women who are underweight benefit from greater weight gain during pregnancy. For women who are overweight and obese, the consequences of weight change during pregnancy are not completely understood. Due to this uncertainty, as a precaution weight loss during pregnancy is not advised[54].

7.4 Micronutrients

Folic Acid

400µg per day[7]

Folic acid is essential for neural tube development. In mPKU 400µg is recommended during preconception and the first trimester in addition to folic acid contained in PS[7]. In some countries 5mg is recommended during pregnancy in obesity or with specific co-morbidities such as diabetes[58, 59].

Vitamin B_{12} status should be monitored to ensure that high intake of folic acid does not mask B_{12} deficiency[7].

Long chain polyunsaturated fatty acids (LCPs)

500mg DHA per day[7]

Pregnant women should receive at least 200mg DHA per day in addition to the 300mg DHA per day recommended for the general population[7, 64].

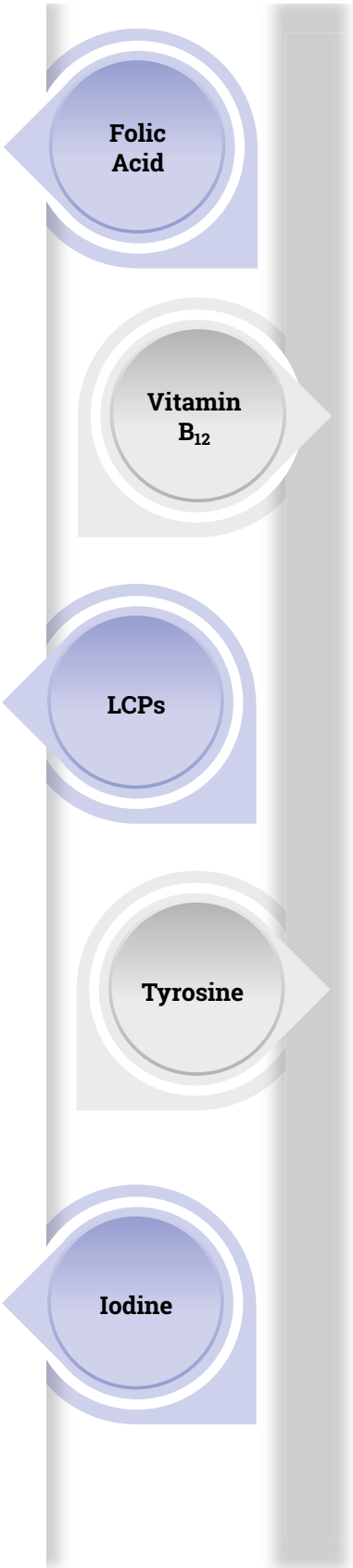
The diet for PKU does not contain dietary sources of omega-3 LCPs. In PKU, DHA can be provided from; fortified PS, prescription products (such as docΩ mega) or over the counter supplements. It is important to check these supplements do not contain vitamin A, fish liver oil, or sources of protein or aspartame.

Iodine

Pregnancy and lactation:
200µg per day[65]

Iodine is an essential nutrient, crucial for neurodevelopment[66].
In the general population there are concerns that women may not consume enough iodine in pregnancy[67,68]. Sources of dietary iodine are severely restricted in the mPKU diet. It was found that average intakes in PKU individuals who were not routinely taking PS was 30µg/day and individuals taking >120% protein recommendations from PS was 120µg/day[61].

Not all PS contain sufficient iodine for pregnancy and lactation. It is important to consider appropriate supplementation to meet increased requirements.



Vitamin B_{12}

Pregnancy: 4.5mg per day[60]

Lactation: 5.0mg per day[60]

Patients with PKU are at risk of vitamin B_{12} deficiency[21, 61-63], the Vitamin B_{12} status (including functional marker plasma homocysteine and/or methylmalonic acid) for all individuals with PKU should be regularly reviewed and corrected if low[7].

In mPKU, insufficient vitamin B_{12} is associated with infant cardiac defects[15].

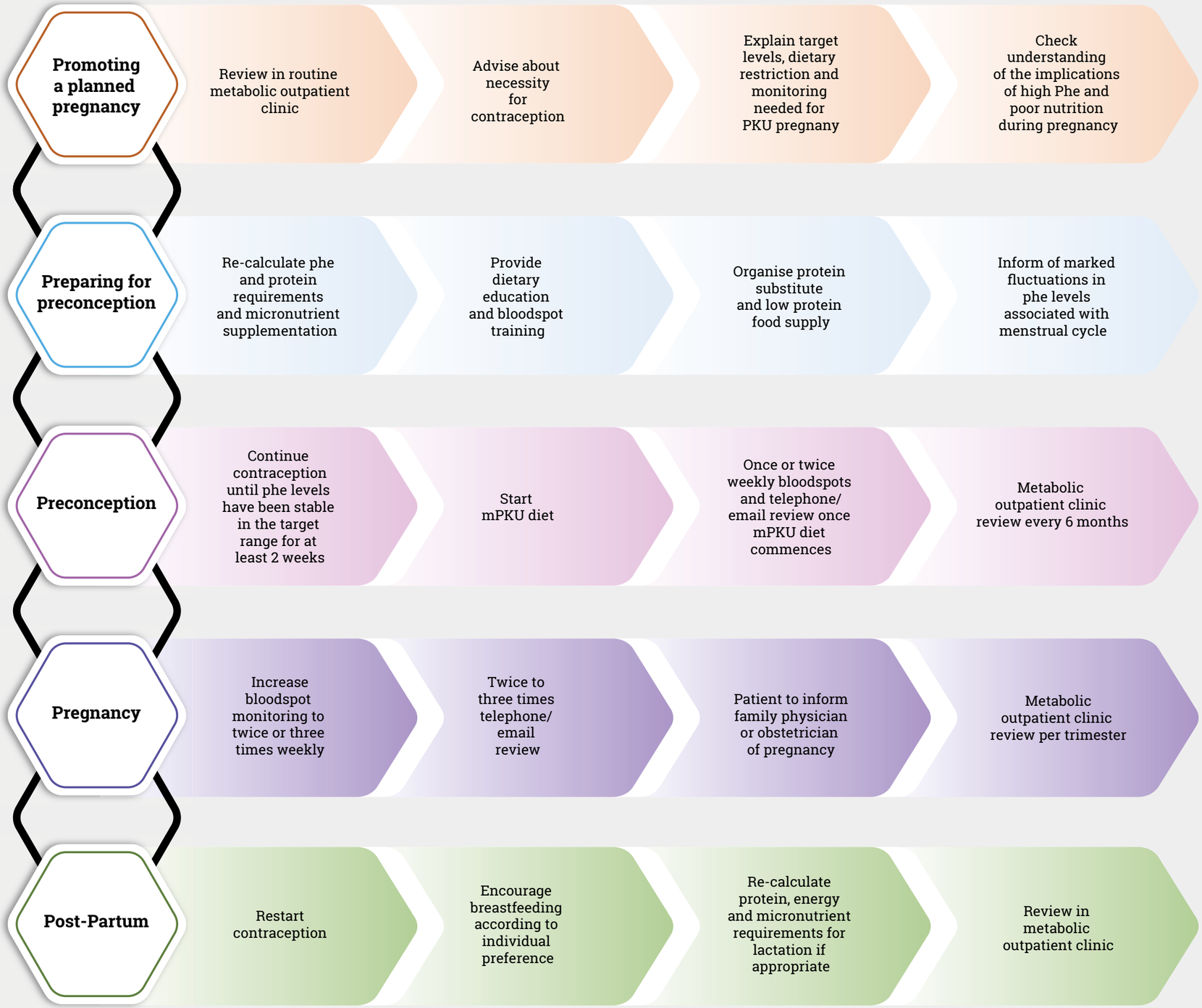
Tyrosine

6g per day[7]

In PKU, tyrosine (tyr) becomes an essential amino acid. At least 6g of tyr is adequate to meet requirements for pregnancy[7].
Sufficient tyr should, therefore, be provided by PS.

Some centres may supplement with additional tyr. Vitaflo's Tyrosine 1000 single dose amino acid sachets are available. These are presented in single dose sachets, making it convenient to add into the diet, if required.

Summary of maternal PKU management



8.0 Vitaflo Range to support the dietary management of mPKU

8.1 Vitaflo's range of protein substitutes (PS)

Designed with features intended to aid adherence, which include non-medical names and packaging, promote improved adherence to facilitate metabolic control.

Product	Presentation	Pack	Flavours	Micronutrients	LCPs	Suitability for mPKU management
PKU cooler 	Ready-to-drink resealable pouches	20g PE 15gPE 10gPE	Red White Orange Purple Yellow	Yes	Yes (DHA)	**A familiar product** and so may be a first choice for the mPKU diet for some. Available in a **range of pack sizes allowing dose adjustment** in 5g PE intervals as nutritional needs change. Smaller pack sizes **can facilitate a 'little and often' approach** to taking PS. Could be useful during nausea as consumed directly from the resealable pouch, limiting exposure to the smell of the product. **It is the only ready-to-drink PS available in a neutral (white) flavour which might be less intense**
PKU Air 	Ready-to-drink resealable pouches	20g PE 15gPE	Red (Berry blast) White (Caribbean Crush) Green (Citrus Twist) Yellow (Mango Breeze) Gold (Coffee fusion)	Yes	Yes (DHA)	Specifically designed with the needs of adults in mind with **flavour blends designed for a mature palate**. This will offer **novel flavour options** to individuals returning to PS for the mPKU diet. **The lowest calorie ready-to-drink PS for PKU**. Could be useful during nausea as consumed directly from the resealable pouch, limiting exposure to the smell of the product.
PKU Express 	Pre-measured, single use sachet	20g PE 15gPE	Unflavoured Orange Lemon Tropical	Yes	No (add docomega for DHA)	Flexible preparation options. **Suitable to be taken as a paste, low volume mini drink or diluted to be a long drink, adapting to changing preference throughout pregnancy.** **Pre-measured, accurate** dose of powdered PS which **can be personalised** by reconstituting with water, protein free milk alternatives or other permitted drinks to make shakes and smoothies. Provides a **lightweight option that is easily transportable** for individuals travelling for work or leisure during pregnancy.
PKU sphere 	Pre-measured, single use sachet	20g PE 15gPE	Red Berry Vanilla	Yes	Yes (DHA)	GMP-based PS provide an alternative taste to traditional amino acid PS which may **optimise adherence**. Similar to the appearance and smell of standard protein shake. Using the PKU sphere shaker, PKU sphere can be **reconstituted on the go without appearing to be a medical product.** **Due to the phe content (36mg phe per 20g PE), caution should be used when incorporating into the mPKU diet.** Changes should be made gradually and closely monitored. It is suggested that during pregnancy dietary phe intake should be adjusted to account for the phe content from GMP-based PS.

* Lower compared to traditional protein substitutes

8.2 Vitaflo's low protein foods

Designed to maximise lifestyle features. All products have non-medical names and packaging, are designed to maximise choice and versatility in recipes and optimise taste. Vitaflo's chefs and dietitians are constantly creating new recipes, many in collaboration with PKU individuals and their metabolic dietitians. These recipes are available on **www.vitafriendspku.co.uk.** For recipes which provide directions on how to add or remove phe exchanges to suit the individual ask your Vitaflo representative for the Live Life Well recipe pack.

Product	Presentation	Description	Suitability for mPKU management
ProZero 	250ml and 1L cartons.	Protein free, liquid blend of carbohydrate. and fat.	A versatile ready-to-drink, protein free alternative to milk. It can be enjoyed on its own, heated, cooked or frozen. Has packaging similar to other commercially available milk alternatives so less likely to draw attention to it being a special dietary product. Useful for adding energy to PS or the LP diet whilst phe tolerance is low.
Mini Crackers 	40g individual	Low protein herb flavour snack crackers.	A ready-to-eat snack that is light weight and portable, fitting easily into a handbag or lunch box. Useful to add energy into the diet. Having non-medical packaging means they can be eaten on the go without drawing attention to a special dietary product.
Vitabite 	25g bars.	Low protein, high energy, chocolate flavoured bar.	A versatile ready-to-eat chocolate flavoured snack. It can be eaten on its own, melted, cooked or frozen. Useful to add energy into the diet. Light weight and portable easily to eat on the go.
Fate special foods Fate SPECIAL FOODS	This versatile range enables cooking of every-day staples, exciting meals and sweet treats which help to normalise and personalise a very strict diet. Fate LP recipes are available in the 'Fate Special Recipes' recipe book, many have video tutorials on the Fate Special Foods YouTube channel which can be found via their website www.fatespecialfoods.com. Many of the Vitaflo recipes also use Fate products.		
	All purpose mix	500g	A versatile mix that can be used for LP bread, pastries, pancakes, scones, muffins and cakes and much more.
	Chocolate cake mix	2 x 250g	Chocolate flavoured mix for light sponge cakes, puddings and crumbles.
	Cake mix	2 x 250g	For light sponge cakes, puddings and crumbles as an all in one mix.

8.3 Vitaflo's additional supplementary products

Product	Presentation	Description	Suitability for mPKU management
docΩmega 	4g sachet = 200mg of DHA.	A powdered blend of Docosahexaenoic Acid (DHA) on a carbohydrate base.	Pre-measured for a simple and accurate dosage of DHA which can be taken on its own, mixed with permitted drinks or added to powdered PS.
Tyrosine1000 	4g sachet = 1000mg of L-Tyrosine	A powdered tyrosine amino acid supplement on a carbohydrate base.	Flavourless powder which can be mixed with water, permitted drinks or added to powdered PS such as PKU express or PKU sphere before reconstituting.

9.0 References

9.0 References

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10.0 Appendices

10.1 Example family physician prescription letter

This is a an family physician prescription request letter which is used at CDMU. This letter is written following the individual's first visit for preconception diet counseling and aims to reinforce the importance of the diet and dietary products to the individual's family physician.

Hospital name and logo

Metabolic Unit, 224 Hospital Lane

Tel: 0984 215 552 Ext.73604/5
Email: metdietitian@hospital.com

Family Physician Address

Date

Dear Dr,

RE: Patient name and address

Diagnosis: Phenylketonuria
Planning a pregnancy

(INSERT PATIENT NAME) was seen in our clinic for review of her Phenylketonuria (PKU) on (INSERT DATE). At that time, the patient and her husband stated that they would like to start a family in 6 months time. Consequently we arranged to see them for dietary education and practical cooking on (INSERT DATE). The patient and her partner have received intensive practical dietary education on how to manage a PKU diet during pregnancy and during the pre-conception period.

The foetal abnormalities associated with untreated or inadequately treated maternal PKU are: congenital heart disease 12%; mental retardation 92%; microcephaly 73%; birth weight <2500g 40% and spontaneous abortion 24%. In order to reduce the risk of these abnormalities, dietary phenylalanine control must start before conception and continue throughout the pregnancy. On (INSERT DATE) her phenylalanine level was ….μmol/L, with the target range for phenylalanine concentrations during pregnancy being (INSERT RANGE) μmol/L.

The diet is immensely demanding and requires a severe restriction of all normal sources of protein. This necessitates careful supplementation with PKU protein substitutes, vitamins, minerals and trace elements. Adequate protein-free calories are provided by the specially manufactured low protein prescription foods.

Her initial dietary regimen will be:
- 8 x 50mg phenylalanine exchanges (~8g natural protein from normal dietary sources)
- 4 x 25g sachets PKU Express / day (a source of phenylalanine-free amino acids as well as vitamins and minerals)
- Various low protein prescription foods – **see list below**

We would appreciate it if you could please prescribe all of the following ACBS approved items:

Product name	Manufacturer	PIP code	Quantity required

We shall see (INSERT PATIENT NAME) in (Metabolic Consultant's name) metabolic clinic on (date) and at regular intervals thereafter.

Between clinics phenylalanine control will be closely monitored by blood tests twice per week (increasing to three times per week when pregnant) and frequent telephone conversations.

Please do not hesitate to contact us if you have any queries regarding (INSERT PATIENT'S NAME)'s dietary management.

With best wishes,

Metabolic Dietitians

Hospital name and logo

10.2 Example monthly summary letter

This is a an example monthly summary letter which is used at CDMU. This letter is sent to the patient, their family physician, midwife (when named) and obstetrician (when named). It aims to provide regular communication on the individual's metabolic control during their pregnancy and provide an opportunity to highlight issues (if any) to all healthcare professionals involved.

Hospital name and logo

Metabolic Unit, 224 Hospital Lane

Tel: 0984 215 552 Ext.73604/5
Email: metdietitian@hospital.com

(PATIENT NAME)

(PATIENT ADDRESS)

date

Dear (PATIENT NAME)

Here is a summary of your results over the past month (INSERT MONTH):

DATE	PHENYLALANINE µmol/L
DD-MM-YY	XX
DD-MM-YY	XX
DD-MM-YY	XX
DD-MM-YY	XX
DD-MM-YY	XX

COMMENT:

Your Phenylalanine levels have been excellent over the past month. The ideal targe range for preconception and pregnancy is (INSERT TAGRET RANGE) µmol/L. Well done and keep up the good work!

DIETARY ADVICE:

Please continue with:

- exchanges per day
- 4 x 25g PKU express /day

NEXT BLOOD TEST: Mondays and Thursdays each week OR Mondays, Thursdays and Saturdays (if pregnant)

Please do not hesitate to contact us if you have any queries.

With best wishes,

Metabolic Dietitians

Copy to: Family Physician

Obstetric team

Hospital name and logo

Notes

Notes

Notes

Notes

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Vitaflo International Ltd,
Suite1.11, South Harrington Building,
182 Sefton Street, Brunswick Business Park,
Liverpool L3 4BQ, UK

+44 (0)151 709 9020
www.vitafloweb.com
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