Returning to Diet

PKU

RETURNING TO A LOW PHENYLALANINE DIET

PHENYLKETONURIA – What is it?

Phenylketonuria (PKU) is a condition inherited from both parents. It was diagnosed when you were about a week old and you were started on the special diet straight away, and is not the result of anything that anyone did wrong during the pregnancy. The problem is that an enzyme in your liver does not work properly. There are hundreds of different enzymes in the body and each one has its own special role in changing one substance into another. The particular enzyme that is faulty in PKU is called Phenylalanine Hydroxylase and is involved in the breakdown of the protein in your diet.

PROTEIN FOODS

The main protein foods are meat, fish, cheese, eggs, soya, tofu and milk and these are needed for growth and maintenance/repair of tissues such as skin, hair etc. Whenever any of these foods are eaten, they are digested and broken down into their smaller building blocks called amino acids. These amino acids are then used for growth and repair.

In any normal diet, there are always huge amounts of all of these individual amino acids which are "left over" after the body has used what has been needed for growth and tissue repair. These surplus amino acids are changed by enzymes into other amino acids.

Phenylalanine → **Tyrosine**

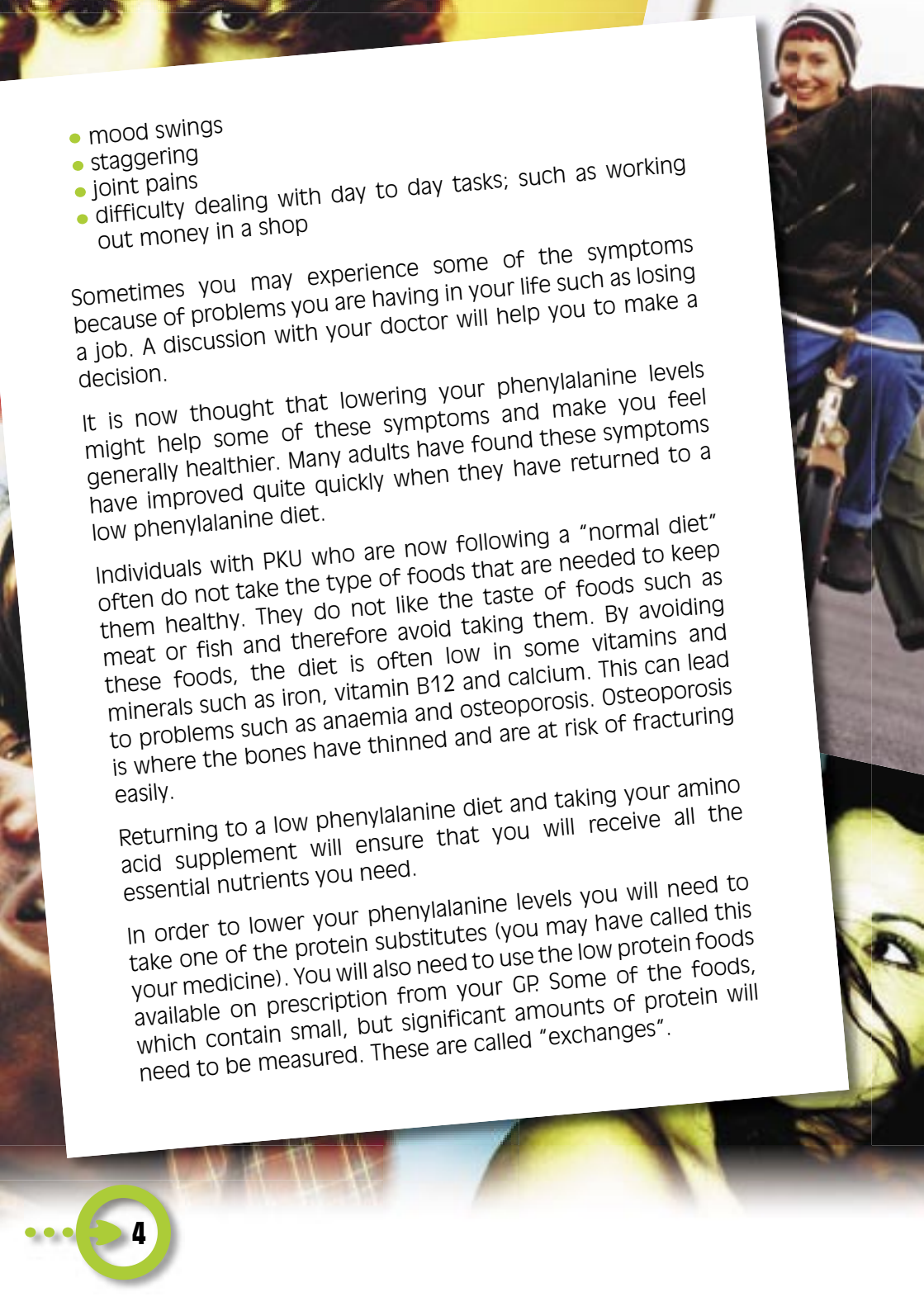
People with PKU have a problem because the enzyme (Phenylalanine Hydroxylase) needed to change the surplus amino acid called phenylalanine into another amino acid called tyrosine is not functioning very well. Although you are able to use phenylalanine quite normally for growth and tissue repair, you cannot deal very well with all the excess phenylalanine, which is surplus to your needs. The result is that the levels of phenylalanine rise very high in your bloodstream. In children, these very high phenylalanine levels prevent the normal development of the brain and nervous system. This is why children with PKU have to follow a strict diet low in phenylalanine to allow them to develop normally. Once the brain had grown and developed to its full extent at around the age of 10 years, the low phenylalanine diet was often discontinued. This is likely to have been what happened with you and you will have been eating a "normal" diet for a number of years.

An important reason to go back to a diet is to plan for a pregnancy. High phenylalanine levels will harm the growing baby and the phenylalanine levels need to be very strictly controlled. You will find the maternal NSPKU leaflet helpful.

WHY YOU MAY WISH TO CONSIDER GOING BACK ON THE DIET?

You may have become aware of a variety of complaints that have been causing you some concern. Some of the symptoms might include:

- poor concentration
- frequent migraine headaches
- tiredness
- anxiety
- skin dryness
- lethargy



- mood swings
- staggering
- joint pains
- difficulty dealing with day to day tasks; such as working out money in a shop

Sometimes you may experience some of the symptoms because of problems you are having in your life such as losing a job. A discussion with your doctor will help you to make a decision.

It is now thought that lowering your phenylalanine levels might help some of these symptoms and make you feel generally healthier. Many adults have found these symptoms have improved quite quickly when they have returned to a low phenylalanine diet.

Individuals with PKU who are now following a "normal diet" often do not take the type of foods that are needed to keep them healthy. They do not like the taste of foods such as meat or fish and therefore avoid taking them. By avoiding these foods, the diet is often low in some vitamins and minerals such as iron, vitamin B12 and calcium. This can lead to problems such as anaemia and osteoporosis. Osteoporosis is where the bones have thinned and are at risk of fracturing easily.

Returning to a low phenylalanine diet and taking your amino acid supplement will ensure that you will receive all the essential nutrients you need.

In order to lower your phenylalanine levels you will need to take one of the protein substitutes (you may have called this your medicine). You will also need to use the low protein foods available on prescription from your GP. Some of the foods, which contain small, but significant amounts of protein will need to be measured. These are called "exchanges".

Generally, your diet should be able to fit in with your lifestyle now you are an adult, although it will involve an amount of planning and work.

- Because the diet is very restricted, it is vitally important that the prescribed amount of protein substitute is taken every day.
- If a vitamin and/or mineral supplement is prescribed it must be taken.
- Blood tests should be sent in at regular intervals so that your blood phenylalanine level can be checked. Adjustments to your diet can then be made depending on the result.

LOW PHENYLALANINE DIET

You may remember the diet being very complicated as often the parents did most of the work and did not teach their children very much about it. This was generally because they had been told that the diet would stop in the early teenage years. It will take you a while to get used to the change from your usual foods to the ones you need to keep the levels of phenylalanine in your blood down. It's probably easiest to think of the diet being in three parts:

1. Foods allowed without counting – free foods

   a) All foods which naturally contain little or no phenylalanine.

   b) Special low protein foods available to you on prescription such as bread, biscuits, pasta. The low protein foods now available on prescription have improved considerably in taste and variety over the years. The NSPKU has produced an Illustrated Guide which has pictures of the many foods available. This will help you choose a good variety of foods to use in your diet.

2. Protein substitute

You need as much protein as any other person. Unfortunately since all protein containing foods such as meat, fish, cheese, eggs etc contain huge amounts of phenylalanine, you are not able to use these. In order to provide you with protein that does not contain phenylalanine, special protein substitutes have been formulated. There are a number of products available either as a drink, tablets, bars or capsules and you can discuss with your dietitian the one that is most suitable for you. It is essential that your protein substitute is taken every day at regular intervals.

3. Phenylalanine food exchanges

The first two parts of the diet do not contain any phenylalanine but remember, you do need to take small amounts of phenylalanine for tissue repair. The phenylalanine food exchanges are designed to provide you with this phenylalanine. Each food exchange contains a standard amount of phenylalanine and by allowing you to eat a certain number of these exchanges every day, your phenylalanine levels will fall. The number of these food exchanges allowed will depend on your blood phenylalanine levels. If your phenylalanine level is too high, the number of exchanges will be reduced and if your phenylalanine level becomes too low, they will be increased.

Sometimes it is possible to lower your phenylalanine levels without strictly counting exchanges, just by being very careful with portion sizes of some protein exchanges. This will be jointly decided between your consultant, dietitian and yourself. This might make the diet easier to manage if you have a busy lifestyle or find counting exchanges makes the diet too difficult to follow.

FOODS ALLOWED

Your dietitian will give you a list of the foods you can safely eat. This list is also available on-line from the NSPKU website www.nspku.org.You will be also be provided with an up-to-date list of all the special low protein foods that are available on prescription from your GP. If you are working, you will probably not qualify for free prescriptions. Since you will be requiring a considerable amount of prescribable items, you should go to your chemist and ask for a pre-paid certificate. The certificate is available for a four or twelve-month period and, for a one-off payment, you will be entitled to all your prescriptions for that period free of charge. The certificate covers all prescriptions including medicines such as antibiotics.

This is the cheapest way of obtaining all your low protein foods and protein substitutes and represents good value for money. A year's supply of dietary foods and low protein substitute would cost a great deal more than the cost of the certificate.

The NSPKU website provides more detailed information about who is entitled to free prescriptions. Ask your dietitian about this if you need help.

BLOOD TESTS

You will remember that a blood test was taken regularly by your parents, but may not remember the numbers that were talked about. It is recommended that you try to keep the phenylalanine levels within the range of 120 – 700 umol/litre which has been shown to reduce the symptoms associated with high phenylalanine levels.

Regular blood tests must be done to monitor the effect of the diet and to let

you know if the number of phenylalanine exchanges that have been prescribed are keeping your phenylalanine levels between the recommended limits. A decision will be made about how often the tests should be taken. If the tests are not carried out then it is difficult to know how effective the diet is at keeping your levels under control.

You may not have taken the blood test by yourself before and you will be shown how to prick your finger and drop the blood on to the card or into the capillary tube if this is used by your clinic. Your doctor and dietician will discuss with you how often the blood tests should be done.

The NSPKU have a leaflet on how to do the blood test.

PHENYLALANINE FOOD EXCHANGES

These are foods which contain small measured amounts of phenylalanine, and the number allowed will depend on the levels of phenylalanine in your blood.

The food exchange system works in the following way:

Each of the following weights of foods contains the same amount of phenylalanine:

80g boiled or mashed potato

45g chips

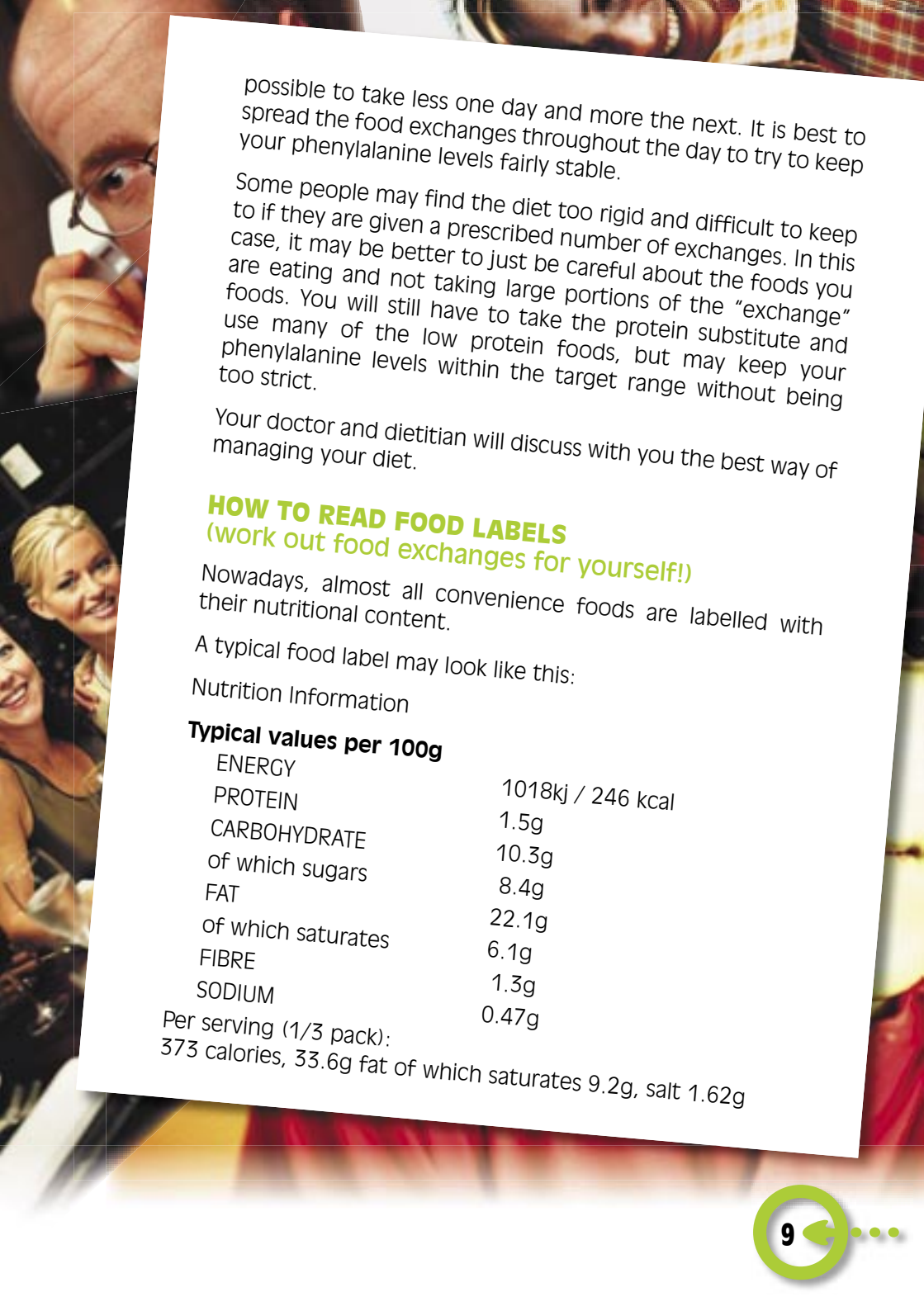
15g cornflakes

20g baked beans

A more extensive food exchange list will be supplied by the dietitian.

This means that you can swap or exchange each of these foods in the weights indicated and you will still be taking the same amount of phenylalanine.

It is important that these are measured carefully and the number allocated should be taken each day i.e. it is not



possible to take less one day and more the next. It is best to spread the food exchanges throughout the day to try to keep your phenylalanine levels fairly stable.

Some people may find the diet too rigid and difficult to keep to if they are given a prescribed number of exchanges. In this case, it may be better to just be careful about the foods you are eating and not taking large portions of the "exchange" foods. You will still have to take the protein substitute and use many of the low protein foods, but may keep your phenylalanine levels within the target range without being too strict.

Your doctor and dietitian will discuss with you the best way of managing your diet.

HOW TO READ FOOD LABELS
(work out food exchanges for yourself!)

Nowadays, almost all convenience foods are labelled with their nutritional content.

A typical food label may look like this:

Nutrition Information

Typical values per 100g

ENERGY	1018kj / 246 kcal
PROTEIN	1.5g
CARBOHYDRATE	10.3g
of which sugars	8.4g
FAT	22.1g
of which saturates	6.1g
FIBRE	1.3g
SODIUM	0.47g

Per serving (1/3 pack):
373 calories, 33.6g fat of which saturates 9.2g, salt 1.62g

This label can be a bit confusing as you have to look at the size of the pack. In this case it is a jar of Korma sauce and there is 455g in the jar. So a 1/3 of the jar is 151g.

To work out how much of the sauce will give you 1g protein or 1 exchange:

Divide 100 by the protein content per 100g

i.e $\frac{100}{1.5} = 66g$

So - 66g of sauce gives 1g protein or 1 exchange

It is usual to round up or down the amount to the nearest 5g. Although a list of food exchanges will be given to you, it is likely you will see some food in the supermarket, which may well be quite suitable for you to have. You can work out how much of the food would be equal to 1 exchange. If you have difficulty doing this then your dietitian will be able to help you.

A quick protein conversion calculator is below:

G of protein / 100g	Amount of food that can be given for 1 exchange
0.3	Free
0.5	200g
1.0	100g
1.5	65g
2.0	50g
2.5	40g
3.0	35g
3.5	30g
4.0	25g
4.5	20g
5.0	20g
5.5	20g
6.0	15g

6.5	15g
7.0	15g
7.5	15g
8.0	10g
8.5	10g
9.0	10g
9.5	10g
10.0	10g

Depending on the type of food, you may find that the quantity for 1 exchange is a very small portion, so you could use 2 or 3 exchanges of the food in this case.

Food labels will also indicate whether or not Aspartame, an artificial sweetener which contains phenylalanine, is contained in the food or drink. Food and drinks containing Aspartame must be avoided.

Should I go back on diet?

It will be up to you whether or not you decide to go back onto the diet. The diet does require some planning and organisation in order to be effective. Your clinic will be able to support you and give you all the information you require to make an informed decision to start the diet.

You should be able to feel the benefits within a very short period of time if you follow the guidance given by your clinic. Keeping the diet going while you go about your daily activities is more difficult and you will need a lot of support from your family, friends and partner, but it is worth it.

For more information look at the NSPKU website:

www.nspku.org

Or Phone : 0845 603 9136 or Email: info@nspku.org

Returning to Diet

P K U

Produced by:

Barbara Cochrane in conjunction with the Medical Advisory Panel of The National Society for Phenylketonuria

NSPKU

The National Society for Phenylketonuria (United Kingdom) Limited

The Society is a registered charity.
It offers support to people with PKU and their families by producing various publications including a quarterly newsletter, organising formal and informal meetings and conferences.

NSPKU, PO Box 26642, London N14 4ZF

Helpline: 020 8364 3010

Email: info@nspku.org

www.nspku.org

Charity No. 273670

Company No. 1256124
July 2005/2,000