PHENYLKETONURIA

(PKU)

A LEAFLET FOR TEACHERS

What is phenylketonuria?

Phenylketonuria (PKU) is a rare, inherited disorder which prevents the normal metabolism of foods containing protein.

Protein, one of the main components of our diet, is made-up of amino acids. In the process of digestion and metabolism dietary protein is broken down into its constituent amino acids and then reassembled into tissue for growth and repair of the body. Phenylalanine is one of the amino acids which make up protein and normally any excess phenylalanine is converted into another amino acid – tyrosine. In PKU this conversion does not take place as normal and the excess phenylalanine accumulates in the blood.

Untreated, this accumulation of phenylalanine impairs brain development and leads to mental retardation.

WITH EFFECTIVE TREATMENT CHILDREN WITH PKU DEVELOP INTO NORMAL HEALTHY CHILDREN.

How is PKU diagnosed?

All babies have a blood sample taken between the 6th and 14th day of life, this is sometimes known as the Guthrie or Scriver test. The sample is used to screen for PKU.

What does the treatment involve?

The treatment involves limiting the amount of phenylalanine in the child's diet. Phenylalanine is not totally excluded as the child requires a small amount for normal growth and development. The balance between excess and adequacy is measured by regular blood tests and the diet may be altered according to the results.

What are the basic principles of the diet?

- Meat, fish, cheese, eggs and milk are rich in protein and therefore phenylalanine, so they are not allowed.

- Other foods which contain some protein such as potato, and cereals are given in small measured quantities so that the blood phenylalanine, which is measured regularly, is kept within safe limits. These measured foods, known as portions or exchanges, are spread out between the day's meals.

- Most fruit, vegetables, salads and also sugar, jam, syrups and fats such as butter, lard and cooking oils can be used quite freely. There are also many special manufactured foods which are low in protein. These include low protein bread, biscuits, flour, spaghetti and other pastas and milk substitutes. They are available on prescription for the child with PKU.

- As the high protein foods such as meat, fish, cheese and eggs cannot be eaten, they have to be replaced by a special protein substitute from which the phenylalanine has been removed. This is given in the form of a drink, paste, capsules or a bar. Vitamins and minerals must also be included in the diet and are given either in the protein substitute or separately. The phenylalanine free protein substitute should, where possible, be taken at mealtimes along with the phenylalanine exhanges. It has a strong unpleasant taste and smell, however negative comments should be avoided.

HOW SHOULD THIS BE MANAGED AT SCHOOL?

Management of PKU involves the concentrated effort, co-operation and understanding of everyone who is involved in the care of the child. The maintenance of such a strict dietary treatment requires discipline and continual encouragement whilst avoiding an over sympathetic or protective attitude towards the child.

School lunches

The child's parents and dietitian will advise on suitable catering arrangements at school. Many families prefer their child to have a packed lunch but it is possible, with appropriate advice, to provide a school lunch. It is important that the remains of packed lunches are sent home so that parents know what has been left and that the child does not swap food with other children as this could lead to incorrect protein being taken. There is a Packed Meal Booklet available from the NSPKU.

Snacks, tuck-shops and sweets

There are a number of sweets allowed freely in the diet and these can be used as snacks. Tuck shops may stock suitable sweets. A piece of fruit also makes a good snack food. If parties or treats are to be given at school it would be advisable to check with the parents and have some suitable foods available for the child with PKU.

It is worth noting that many foods & drinks contain the sweetener Aspartame which is unsuitable for children with PKU as it contains phenylalanine.

Dietary Compliance

If a child with PKU eats 'forbidden foods' there will not be any noticeable ill-effects. You should be reassured that the occasional dietary indiscretion will not cause any lasting harm, neither will they have any violent reaction.

However such incidents should not be ignored as they may indicate or lead to more frequent lapses of diet. Prolonged or frequent dietary indiscretion could have an adverse effect on the child. The diet can be stressful to both child and parents. Non PKU children should be discouraged from eating the special low protein foods, however if they do they will not come to any harm.

Educational Achievement

Children with PKU should be treated in the same way as all other children in your class. Early diagnosis and careful management of the condition results in normal healthy children who should aim for the same educational goals as their classmates. Children with PKU have regular developmental assessments. No additional assessments should be required.

Produced by the National Society for Phenylketonuria
and its Medical Advisory Panel.

NSPKU

The National Society for Phenylketonuria (United Kingdom) Limited

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

Further information and details can be obtained by contacting:
The NSPKU Helpline on: 0845 603 9136
or writing to:
The National Society for Phenylketonuria (United Kingdom) Ltd.
7, Lingley Lane, Wardley, Gateshead, Tyne & Wear, NE10 8BR

Charity No. 273670 Email: nspku@ukonline.co.uk Company No. 1256124

http://web.ukonline.co.uk/nspku July 1999/1200