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This brief section in English language introduces into PKU and its special dietary needs. It mainly concerns the medical community.

Section 2: E.S.PKU Member States and Their Languages

- In the first column, the E.S.PKU Member States are listed in their English version in alphabetical order;
- The second column, indicating the language(s) spoken, leads directly to the main section 3:

Section 3: How to Explain PKU in Another Language

- The explanations on PKU are listed in alphabetical order of the English version of the languages;
- Each language is printed on a separate page.

If someone is asking for a travel information for one of the countries, you have nothing to do but to copy:

Section 1: “General Information on Phenylketonuria” for the medical community in other countries;

Section 3: The page with the information on PKU in the national language requested;

In case no information in one of the national languages is available, please copy in section 3 the English version.

If you have any additional question, such as addresses of metabolic centres in a particular country, please contact the E.S.PKU Representatives. Their names and addresses are listed in the “E.S.PKU Handbook”, Appendix 2 or in the E.S.PKU Web Sites: <http://www.ping.be./ESPKU>

Section 1: General Information on Phenylketonuria

Phenylketonuria (= PKU) is one of the inborn errors of amino acid metabolism. It is a recessively inherited disorder, which concerns the enzyme phenylalanine hydroxylase, located in the liver. This enzyme normally converts the essential amino acid phenylalanine, one of the building blocks of proteins, to tyrosine.

In case of PKU - due to the partial or complete lack of enzyme activity - phenylalanine is not converted, but accumulates in blood and tissues. In untreated PKU, retarded intellectual development is the most important and constant clinical feature. Therefore, early treatment is imperative. Children with PKU detected by routine neonatal screening and beginning of treatment soon after birth generally develop within the normal range.

So far, treatment for PKU only consists of a diet low in phenylalanine and individually "tailored" to control blood phenylalanine levels in a "safe" range. Longterm studies have shown that this treatment is effective but, in order to prevent brain damage, has to be followed strictly.

The basic principles of the "PKU diet" are as follows:

- The diet has to be calculated according to age for optimal intake of energy and nutrients;
- Most of the natural sources of proteins - such as meat, milk, milk products, eggs, bread, pasta, and pulse - have to be replaced by an amino acid mixture, which is free of phenylalanine;
- The so-called "phenylalanine requirement" for growth and replacement of body proteins has to be covered by some foods with a low content of protein and therefore of phenylalanine: some vegetables, fruit and specially manufactured "dietetic foods", such as bread, cakes, and pasta.

Based on these "columns", PKU patients have their individually "tailored" diet plans to follow. In case of an illness, such as a cold or diarrhoea and in an accident, a rapid deterioration of phenylalanine metabolism can occur. Therefore, an immediate treatment in an experienced centre, usually a nearby university children's hospital, is required.

The initial steps taken with patients affected by other disorders of amino acid metabolism, such as organic acidemias and urea cycle defects, are even more crucial to the clinical course. Those patients have also immediately to be admitted to an experienced children's hospital.

Section 3: How to Explain PKU in Another Language

Danish

Af helbredsmæssige årsager må jeg ikke spise madvarer, der indeholder meget protein, f.eks. kød, fisk, æg, ost og soyaprodukter. Grøntsager, såsom kartofler, majs, ærter, og hovedkål samt morgenmadsprodukter, kan jeg kun spise i begrænset mængde. Frugt, bladsalat, sukker, syltetøj, olie, smør og margarine kan jeg spise i normale mængder. Fødemidler sødet med Aspartame er ikke tilladt.

For at jeg skal føle mig helt rask et jeg nødsaget til at overholde denne strenge diæt samtidig med at jeg skal indtage en speciel aminosyreblending.

Nogle få sætninger der kan hjælpe dig: Jeg er på streng diæt

1. Kunne jeg få en tallerken med kun kartofler ?

Indeholder dette kød/fisk/æg/ost

2. Kunne jeg få nogle kartofler/chips/pommes frites sammen med grøntsagerne ?

3. Må jeg bede om noget frugt ?

4. Kunne De koge dette pasta til min aftensmad ?

5. Kunne De riste dette specielle brød for mig ?

6. Kunne jeg få lov til at låne en vægt til min diæt-mad ?

7. Jeg kan ikke tåle mad eller drikke, der indeholder **ASPARTAME**

I specielle tilfælde kontakt venligst et af behandlingsstederne.

Notes for my Travel:

Section 3: How to Explain PKU in Another Language

Dutch

Apart from The Netherlands, Dutch is also spoken in parts of Belgium.

Omwille van gezondheidsredenen mag ik geen eiwitrijke produkten eten zoals vlees, kip, vis, eieren, kaas, melkprodukten, 'gewoon' brood en soja. Groenten zoals aardappelen, maïs, kool en granen zijn slechts in beperkte en afgemeten hoeveelheden toegelaten. Fruit, bladgroenten, suiker, confituur, olie, boter en margarine zijn toegelaten in normale hoeveelheden. De voedingsmiddelen, gezoet met Aspartaam zijn verboden. Om mij in goede gezondheid te houden, ben ik verplicht een strikt dieet te volgen aangevuld met medische voedingsmiddelen.

Hier volgen enkele zinnen die u kunnen helpen: Ik volg een strikt dieet

1. Kunt u mij een bord met uitsluitend groenten brengen ?
Bevat deze schotel vlees, kip, vis, eieren, kaas, melkprodukten ?
2. Kan ik enkele aardappelen/ chips/ frieten bij de groenten krijgen?
3. Kan ik wat fruit krijgen ?
4. Wilt u voor mij (eiwitarme) pasta koken als avondmaal ?
5. Wilt u voor mij dit speciale (eiwitarm) brood roosteren ?
6. Kunt u mij een weegschaal geven om de poeders (medicatie) af te wegen ?
7. Ik mag geen voedsel of drank gebruiken die ASPARTAAM bevat.

In geval van nood, kunt u onmiddellijk contact nemen met het dichtsbijzijnde medisch centrum?

Notes for my Travel:

Section 3: How to Explain PKU in Another Language

English

For medical reasons I am not allowed to eat any products which contain a lot of protein. i.e. meat, fish, eggs, cheese and soya. Vegetables like potatoes, sweet corn, cabbage leaves, pulses and cereals are allowed in limited quantities and have to be measured exactly. Fruit, leaf vegetables, sugar, marmalade, oil, butter and margarine are permitted freely. Any foods, drinks or medications sweetened with Aspartame, are not allowed.

To stay in good health, I have to follow this strict diet with medication.

A few questions to help you: This strict diet is vital to keep me in good health.

1. Can you bring me a plate of vegetables only?
Does this dish contain meat/fish/egg/milk/cheese?
2. Please, could I have some potatoes/chips/french fries potatoes with the vegetables?
3. Please, may I have some fruit?
4. Can you cook this pasta for my meal tonight?
5. Can you toast this special bread for me?
6. Can you bring me a weighing scale to prepare my medical food?
7. I cannot take food or drinks containing ASPARTAME

For any urgent need, please contact the nearest medical centre.

Notes for my Travel:

