Which restricted diet shall we propose in PKU as we approach the year 2000?

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The use of a low-Phe diet has certainly completely modified the neurological outcome of patients with PKU. Under treatment, these patients do not display the severe mental retardation and neurological impairment found in older PKU patients born more than 40 years ago.

Treatment consists of a semisynthetic diet of foods naturally low in Phe, together with 'protein-free' products (the so-called 'caloric module') and supplemented by Phe-free protein supplement (the 'nitrogen module').

The day-to-day use of this modified diet is, however, very frustrating for the patient, leading to poor compliance in adolescence or adulthood. In addition, due to the very bad taste of the product, patients refrain from using the nitrogen module correctly. In developing countries, the priority of the health policy is treatment of infectious diseases and little of the budget is left for the treatment of metabolic diseases. The very expensive amino acid mixture currently used in Western countries is virtually unavailable in developing countries, leading to very bad dietary and metabolic control in PKU patients living there.

Recently, J. Laloux, a PhD student in our department, had the opportunity to propose and study a new approach in the use of Phe-free protein substitute. It is already known that some peptides obtained after hydrolysis of cows' milk are low in Phe residues. The difficulty remains to separate these from the other peptides. Different methods have been tested where the Phe content was reduced from 5%, as in the cows' milk protein, to less than 0.1%. The taste of these peptides (which range from 13 to 32 amino acids residues) is neutral and, added to natural food, does not modify the original taste.

There are some differences between methods in terms of purity, yield and costs of the procedure. For example, with the alcohol precipitation as a one-step procedure, we obtained a product where the Phe content was reduced by 80%. This is not sufficient in classical PKU where the oral Phe tolerance is less than 350 mg/day. However, in the southern part of Europe, this may be different as it is known that, in countries around the Mediterranean, the incidence of atypical or mild PKU with higher tolerance is much more frequent than in Northern Europe. One should ask whether this inexpensive and partially purified peptide should not be proposed to replace the amino acid mixture in countries where these products are not available due to their high cost. In addition, the use of peptides instead of a mixture of free amino acids could be of better nutritional value.

It should be mentioned here that no clinical study has been conducted so far and no final conclusion can be drawn at the present time.