

DIETARY THERAPY FOR PHENYLKETONURIC PATIENTS IN TAIWAN

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Phenylketonuria (PKU) is an inherited metabolic disease. The patients will be mentally retarded, if they are not diagnosed and treated early enough. In order to detect affected babies and start the treatment within 2 months of age, mass neonatal screening programs have been organized in many developed countries. Normal mental development occurred in classical phenylketonurics, who were treated by dietary management until the age of 6-10 years old. In the past 2 years, 7 cases of classical PKU and 4 cases of hyperphenylalaninemia virants (HPV) have been diagnosed in the pediatric clinic, in mentally handicapped school children and in mental institutions in Taiwan by our laboratory. In order to establish an adequate dietary management system for Chinese phenylketonuric patients, 9 of them (5 PKU and 4 HPV) were put on a dietary therapy trial with special formula and local foods. The serum phenylalanine level was maintained between 2 to 8 mg/dl for patients younger than 10 years old. Because dietary control may not improve intelligence for older children and the dietary control of early

diagnosed patient in the future will be relaxed at age of 10, the serum phenylalanine level was controlled below 20 mg/dl for patients over 10 years old in this study. Preliminary results show that the serum phenylalanine was dramatically decreased in response to the dietary control. Both the physical and mental development of the restrictively controlled patients and the behavior of the loosely controlled patients improved. But the definite developmental effect of this dietary management method still requires long term evaluation. There are large individual differences in phenylalanine tolerance. For HPV, the daily phenylalanine intake might be 80-150 mg/kg. But the classical PKU can only tolerate 18-30 mg/kg daily phenylalanine intake. These data indicate that individualized management with close serum phenylalanine monitoring is essential to an adequate dietary therapy. The dietary management method and the phenylalanine equivalency table of local food used in this study should be applicable to future phenylketonurics discovered by a neonatal screening program in Taiwan.

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