News
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June 7, 1985

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****Update on PKU for ex-patients and their families scheduled for June 15

DALLAS--A team of health personnel at The University of Texas Health Science Center at Dallas is looking for families in which at least one member is known to have PKU or be a PKU carrier. PKU, short for phenylketonuria, is a treatable inherited disorder, and much new information has been learned since it was first discovered about 50 years ago.

Medical, nutritional and social-work professionals are trying to reach families to let them know about a free PKU workshop to be held Saturday, June 15 from 8:45 a.m. to 12:30 p.m. at Children's Medical Center in Dallas. Sessions will be held for adolescents with PKU and for their parents.

PKU is an inherited disorder that can be helped, says Carol Hickey, instructor in Clinical Pediatrics and Nutrition and Dietetics at UTHSCD. Between 25 to 30 years ago, says Hickey, researchers learned that a very low-phenylalanine diet in children can make the difference between a child's healthy development and that child's growing up with possible mental retardation, seizure disorders or behavioral problems.

A child born with PKU has a genetic disorder caused by a defect in a specific protein, an enzyme called phenylalanine hydroxylase, that is needed to convert phenylalanine to another amino acid called tyrosine. This reaction, which takes place under normal conditions, is necessary to use the protein in food. In the child with PKU, there is no such chemical reaction; therefore, these children have a build-up of phenylalanine in the blood and tissues. In Texas, as in many other states, all new-borns are checked for PKU and parents alerted early so they can receive special medical attention for their children.

Unfortunately, says Hickey, many children who were put on the PKU diet were lost to follow-up after their diet was discontinued between the ages of six and eight years. Children were treated for the first few years of their lives, and everyone assumed that the problem was taken care of because the major part of brain growth was completed at that time. However, today there are speculations that it would be better for children to stay on the PKU diet longer and that some adolescents might benefit from treatment. Furthermore, there has been a great improvement in the diet products.

There is also the question of treatment for unborn children. Because PKU is the kind of genetic disease called an autosomal recessive disorder, parents who do not have clinical symptoms of PKU may be carriers and pass on the disorder. Thus, PKU may be passed down "silently" through both families until two carriers marry and have a PKU baby. New genetic information about the disease is now available to PKU patients.

More importantly, it is now thought that putting a woman who has PKU back on the diet when she decides to have children--preferably *before* she conceives--may offset or prevent mental retardation and other birth defects in her child, which can be caused by the PKU woman's high blood levels of phenylalanine.

Because there is new knowledge in the areas of treatment and diagnosis, The University of Texas Health Science Center at Dallas, its University Affiliated Center and Children's Medical Center are inviting family groups with concerns about PKU to the PKU workshop. The program will include information about the disorder itself, how family planning relates to PKU, information about the PKU diet, diagnosis, carrier testing, vocational and educational issues and special information for adolescents.

(More)

"Many of these decisions--including those about family planning--should be made with freedom of choice after people receive proper education," says Barbara Cambridge, research associate professor and director of social services for the Department of Obstetrics and Gynecology at the health science center. Cambridge counsels young people and their families about PKU-related problems on a regular basis and is a member of the PKU health team.

Also working with PKU patients for over 15 years is Dr. Charles Mize, associate professor of Pediatrics and assistant professor of Biochemistry, who holds both M.D. and Ph.D. degrees. Mize stresses how important it is for both children and their families who have not stayed in contact with medical experts in PKU to learn about recent developments in knowledge about the disorder. Mize regularly sees PKU patients at Children's Medical Center.

Dr. George Wendel, assistant professor of Obstetrics and Gynecology, will work with pregnant women--or women who plan to become pregnant--who want to be closely monitored during their pregnancies.

Other team members participating in the free program include Karen Heller, Cindy Cunningham, Dr. Janice Nici and Melissa Brown.

To get more information or to register for the program, call Carol Hickey at the University Affiliated Center, 214/688-2883.

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