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Maternal PKU—A Continuing Problem

An article in this issue of the Journal, entitled "The New England Maternal PKU Project", by Waisbren, *et al*,[1] calls attention to the crisis facing thousands of young women in the world today that was produced—ironically—by the great success of newborn screening for PKU (phenylketonuria) carried out during the past two decades. These women are in three groups:

- those identified by neonatal screening and treated early, but usually lost to follow-up at 5-8 years of age;
- those born before newborn screening, usually treated late for a few years and then also lost to follow-up;
- those never identified, most often because their phenylalanine (phe) blood level was not considered high enough to warrant treatment ("hyperphe").

Waisman and colleagues first showed the teratogenic effects of elevated phe during pregnancy in the rhesus monkey and also that phe elevation was greater on the fetal side of the placenta.[2] Dent first pointed out the phenomenon in the human species.[3] MacCready and Levy reviewed the problem and initiated a pilot program aimed at detection of PKU on the first premarital examination or prenatal visit.[4]

Lehnke and Levy[5] published a comprehensive review, and Levy organized the New England Maternal PKU Project described in this issue. A national attempt in Canada to address this problem by developing a registry to identify and track all individuals with elevations in phe was described in this Journal[6] and was the subject of an accompanying editorial.[7] The New England project then became part of the Maternal PKU Collaborative Study (for the USA and Canada) organized by Koch and his colleagues.[8] This study is designed to identify, counsel, and treat during pregnancy young women with PKU or any degree of hyperphe.

As is pointed out in the article in this issue,[1] it is of critical importance to initiate dietary treatment *before* pregnancy. All women with any degree of phenylalanine elevation, and who may become sexually active, must be identified and put into a system that includes dietary support, psychosocial support, and metabolic monitoring. This paper describes a method for identifying and tracking these young women, a very necessary first step in effectively treating maternal PKU.

PKU, although rare, has played a major role in our attempt to increase understanding of the nature of man, through research into function of the brain and central nervous system. The attack on maternal PKU is an important part of this process. In the past, most studies dealt with the developing brain, now, because of maternal PKU, with pathogenesis in the fetus. In the future we will have to deal with the effects of elevations in phe on behavior, as was illustrated in Bickel's first publication on the effect of dietary treatment 35 years ago.[9] We will then have to organize new programs to include young men as well as women.

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