PREVALENCE OF HYPOPIGMENTED MACULES

The prevalence of hypopigmented macules among 423 white individuals and their significance in the diagnosis of tuberous sclerosis (TS) were evaluated at the University of Washington School of Medicine and Children's Hospital, Seattle, WA. Twenty (4.7%) had at least one macule, 4 of these had more than one macule, and none had more than three. Two (8%) of 25 hypopigmented macules were identified only with a Wood lamp. Of the 20 individuals with macules, 13 had ophthalmoscopic exams and none showed retinal changes of tuberous sclerosis. A few hypopigmented macules on the skin of otherwise healthy individuals without a family history of TS do not warrant a search for other signs of the disorder. (Vanderhooft SL, Francis JS, Pagon RA, Smith LT, Sybert VP. Prevalence of hypopigmented macules in a healthy population. <u>Pediatr</u> Sept 1996;129:355-61). (Reprints: Sheryll L Vanderhooft MD, University of Utah Health Sciences Center, Department of Dermatology, 50 North Medical Dr, Sat Lake City, UT 84132).

COMMENT. Hypopigmented macules are apparently more common than previously determined, occurring in close to 5% of the general population under 45 years of age. The presence of one to three macules is not by itself a significant risk factor for tuberous sclerosis. As a secondary feature of TS, at least one other manifestation must be present to establish a diagnosis.

MYOCLONUS IN ANGELMAN SYNDROME

A clinical and electroencephalographic study of 11 unrelated patients with Angelman syndrome (AS), confirmed by genetic analysis, is reported from the University of Pisa, Italy. All patients showed the jerky, tremulous, or dystonic motor pattern typical of AS. Using long-term video-EEG and polygraphic monitoring, these abnormal movements were shown to be a form of fast-bursting cortical myoclonus. Antimyoclonic treatment with piracetam in 5 patients produced a marked functional improvement. (Guerrini R, De Lorey TM, Bonanni P, et al. Cortical myoclonus in Angelman syndrome. <u>Ann</u> <u>Neurol</u> July 1996;40:39-48). (Respond: Dr Guerrini, Institute of Child Neurology and Psychiatry, University of Pisa, Via dei Giacinti 2, 56018 Calambrone, Pisa, Italy).

COMMENT. The diagnostic features of Angelman syndrome include ataxia, developmental delay, paroxysmal laughter, microcephaly, and seizures. The "puppetlike" movement disorder is related to a cortical myoclonus.

TOXIC FACTORS IN ATTENTION DEFICIT

POLYCHLORINATED BIPHENYLS AND ATTENTION DEFICITS

The effects of in utero exposure to polychlorinated biphenyls (PCBs) on cognitive function in 212 children at 11 years of age were tested at Wayne State University, Detroit, MI. Prenatal exposure to PCBs from maternal ingestion of contaminated Lake Michigan fish was associated with significantly lower full-scale and verbal IQ scores. Concentrations of PCBs in maternal serum and milk at delivery, only slightly higher than in the general population, caused long-term intellectual impairment, especially affecting memory, attention, and reading comprehension. (Jacobson JL, Jacobson SW. Intellectual impairment in children exposed to polychlorinated biphenyls in utero. <u>N Eng J Med</u> Sept 12 1996;335:783-9). (Reprints: Dr Joseph L Jacobson, Department of Psychology, Wayne State University, Detroit, MI 48202). COMMENT. Deficits in short-term memory and developmental delays, previously noted in infants and at 4 years of age in children exposed to PCBs in utero, have now been demonstrated in children tested at 11 years of age. PCBs may have a long-term adverse effect on cognitive function, and prenatal exposure to these environmental toxins should be included among potential causes of attention deficit disorders in children.

PREDICTIVE VALUE OF LEAD SCREENING PRACTICES

The prevalence of elevated blood lead levels and the accuracy of a lead screening questionnaire in an urban pediatric population were evaluated in the primary care clinics of 10 community health centers in the city and county of Denver, CO. Of approx 3000 low-income children tested, only 0.3% had blood lead levels >20 mcg/dL. The predictive value of the CDC questionnaire was 3%, little better than chance, and the cost of identifying a child with a lead level > 20 mcg/dL was approx \$5000. (France EK, Gitterman BA, Melinkovich P, Wright RA. The accuracy of a lead questionnaire in predicting elevated pediatric blood lead levels. <u>Arch Pediatr Adolesc Med</u> Sept 1996;150:958-963). (Respond: Eric K France MDCM, Department of Preventive Medicine, Kaiser Permanent, 10350 E Dakota Ave, Denver, CO 80231).

COMMENT. The Editor, Dr Catherine D DeAngelis, notes that "when a survey questionnaire is only slightly better than chance, it's better to take the chance and save the money for the blood tests." The Denver Health and Hospitals chose to screen all low-income children between 12 and 30 months of age and to forgo the use of the questionnaire. Universal screening may be omitted in low-prevalence communities, according to current CDC guidelines.

A recent survey of a nationally representative sample of pediatricians found that 53% screen all their patients aged 9 to 36 months, 96% using a blood lead assay. Most of the remainder report screening of high-risk patients only. (Campbell JR, Schaffer SJ, Szilagyi PG et al. Blood lead screening practices among US pediatricians. <u>Pediatrics</u> Sept 1996;98:372-377). In pediatric neurology practice, a blood lead level may be indicated in high-risk children who present with neurodevelopmental delay, ADHD, seizures, or signs of encephalopathy or neuropathy.

CARBAMAZEPINE VS VALPROATE AND COGNITIVE FUNCTION

Effects of carbamazepine vs valproate on cognitive functioning in patients with previously unmedicated epilepsy were evaluated in a prospective, randomized, double-blind Veterans Affairs multicenter study. Patients with seizures showed deficits relative to a normal control group prior to AED therapy. No significant decline from baseline levels of neuropsychological performance was detected over 6- or 12-month treatment intervals for either drug. Patients with high serum VPA levels (mean, 94 mcg/mL) performed less well than controls on measures of concentration and memory. Subtle compromises of cognitive functioning following treatment with VPA or CBZ were suggested by absence of practice effects. (Prevey ML, Delaney RC, Cramer JA et al. Effect of valproate on cognitive functioning. Comparison with carbamazepine. <u>Arch Neurol</u> Oct 1996;53:1008-1016). (Reprints: Mary L Prevey PhD, Neurology 127, VA Medical Center, West Haven, CT 06516).

COMMENT. Carbamazepine and valproate monotherapies have subtle effects on cognitive functioning, when studies are well controlled.