biotinidase deficiency was suspected and confirmed. Both infants responded promptly to biotin, and blood pH and bicarbonate became normal within hours. At 5-11 month follow-up, seizures had not recurred, the EEG and neurologic examinations were normal, but the developmental mental scores on the Bayley Scale were severely retarded. (Kalayci O et al. Infantile spasms as the initial symptom of biotinidase deficiency. J Pediatr Jan 1994;124:103-4). (Reprints: Omer Kalayci MD, Bahcelievler 39, sokak 12/6, 06500 Ankara, Turkey).

COMMENT. Biotin responsive late onset multiple carboxylase deficiency is an autosomal recessive inherited disorder manifested by seizures, alopecia, skin rash, hypotonia, ataxia, hearing loss, and developmental retardation. Lactic acidosis and organic aciduria may be delayed. If untreated the symptoms become progressively worse and coma and death may occur. Symptoms respond rapidly to biotin 5-10 mg daily, but neurologic damage may be irreversible. (Progress in Pediatric Neurology. Chicago, PNB Publishers, 1991, pp547-550). Biotin deficiency should be considered as a possible etiology of infantile spasms. A therapeutic trial of biotin has been recommended in all drug resistant infantile seizures, pending the results of enzyme and metabolic tests. (Ped Neur Briefs Nov 1989).

Infantile spasms or myoclonic seizures were present in 16% of 30 infants with biotinidase deficiency reported from the Medical College of Virginia, Richmond, VA. (Salbert BA, Wolf B et al. Neurology 1993;43:1351). The authors advocated neonatal mass screening for early diagnosis and avoidance of neurologic damage.(Ped Neur Briefs July 1993;7:51).

TOXIC DISORDERS

TOLUENE EMBRYOPATHY

The clinical manifestations of toluene embryopathy in 18 infants with a history of in utero exposure are reported from the Department of Pediatrics, University of Arizona College of Medicine, Tucson, and Maricopa Medical Center, Phoenix, AZ. Mothers were regular abusers of solvents and the fetus was exposed to toluene by maternal spray paint sniffing. Nine of the infants had been exposed to alcohol in addition, but except for an increased incidence of prenatal microcephaly, the resultant phenotype was unchanged. Premature birth occurred in 39%, and 9% died, 54% were small for gestational age, 52% had postnatal growth deficiency, 33% prenatal microcephaly, 67% postnatal microcephaly, 80% developmental delay, and 83% had craniofacial features similar to the fetal alcohol syndrome. Micrognathia, small palpebral fissures, and abnormal ears were most frequent with toluene, whereas the thin upper lip, smooth philtrum, and small nose were more common with alcohol exposure. Other less prominent features common to both toluene and alcohol embryopathies were nail hypoplasia, abnormal muscle tone, hemangiomata, renal anomalies, and altered palmar creases. (Pearson MA et al. Toluene embryopathy: Delineation of the phenotype and comparison with fetal alcohol syndrome. Pediatrics Feb 1994;93:211-215). (Reprints: H Eugene Hoyme MD, Section of Genetics, Dept of Pediatrics, Arizona Health Sciences Center, Tucson, AZ 85724).

COMMENT. It is estimated that 3 to 4% of teenagers engage in paint or glue sniffing. Toluene is the active organic solvent. It is an

underrecognized form of substance abuse with serious acute and chronic toxicities. Acute symptoms of toluene toxicity include dizziness, euphoria, headache, vomiting, vertigo, convulsions, and loss of consciousness, sometimes preceded by delirium. Chronic toluene abuse causes headache, muscle weakness, peripheral neuropathy, nervousness, anemia, petechiae, abnormal bleeding, bone marrow aplasia, irreversible encephalopathy, and renal tubular acidosis. Following an initial report of a "Fetal solvents syndrome"(Toutant C, Lippman S. Lancet 1979;1:1356), the effects of toluene on the fetus have been described infrequently. This Arizona University study of a fetal toluene syndrome compares findings in their 18 patients with others in the literature and with the fetal alcohol syndrome. The authors conclude that the craniofacial teratogenetic effects of toluene and alcohol have a common mechanism.

An additional 6 cases of toluene embryopathy are reported from the Denver General Hospital, Colorado (Arnold GL et al. Pediatrics Feb 1994;93:216). Only one was exposed to alcohol as well as toluene.

LEAD POISONING RISK ASSESSMENT: BLOOD LEAD SCREENING

The Centers for Disease Control and Prevention (CDC) lowered the blood lead level (BPb) considered a toxic risk to children from 25 mcg/dL to 10 mcg/dL in Oct 1991. A five-item questionnaire is now completed at regular office visits for all children from 6 months to 6 years of age to identify those at high risk of lead exposure. In some States, including Illinois, all children are required to have BPb testing at 12 months of age or before entering a state-licensed day care, preschool, or kindergarten. High risk infants must be tested at 6 months and biannually.

A study to determine the efficacy of the questionnaire and prevalence of elevated BPb in 1393 suburban children at 12 and 24 months of age is reported from the Department of Pediatrics, Children's Memorial Hospital, Northwestern University, Chicago, Illinois. A venous BPb =/> 10 mcg/dL was found in 2.1%, and none was >30 mcg/dL. CDC and Illinois screening tests failed to predict high risk exposure in 9 of 29 (31%) children with elevated BPb. Living in a pre-1960 house, a question not included in CDC or Illinois screening, was most predictive, with positive correlation in 24 of 29 (83%) children with elevated BPb levels. The authors point out that optimal risk assessment questions may vary in different areas and populations. (Binns HJ et al. Is there lead in the suburbs? Risk assessment in Chicago suburban pediatric practices. Pediatrics Feb 1994;93164-171). (Reprints: Helen J Binns MD, Children's Memorial Hospital, 2300 Children's Plaza, Chicago, IL 60614).

COMMENT. Questions about the home environment were the most sensitive indicators of elevated lead levels in a similar study reported from the California Pacific Medical Center, San Francisco, CA. (Tejeda DM et al. Do questions about lead exposure predict elevated lead levels? Pediatrics Feb 1994;93:192-194). An abbreviated screening using only the first three items was as effective as the complete CDC questionnaire in a study at the University of Rochester, NY.(Schaffer SJ et al. Lead poisoning risk determination in an urban population through the use of a standardized questionnaire. Pediatrics Feb 1994;93:159-163). Selective screening with a community-specific questionnaire is proposed following a study at the Gunderson Clinic, La Crosse, WI, which found a great variability in prevalence of elevated BPb between clinics