SEIZURE DISORDERS

SHIGELLOSIS FEBRILE STATUS EPILEPTICUS

A 4-year-old boy who became blind, deaf and mute after status epilepticus caused by hyperpyrexia from shigellosis is reported from the Sophia Children's Hospital, Rotterdam, The Netherlands. Hyperpyrexia and diarrhea developed 2 days after eating tainted Chinese food at a family feast. Stool cultures grew *Shigella flexneri.* CT showed cerebral swelling. He had several generalized tonic clonic seizures followed by status and prolonged coma. On day 9 he opened his eyes and localized painful stimuli. He was blind, deaf and mute. Vision and hearing recovered within 6 months but expressive language impairment was more persistent. At 4 year follow-up he could repeat simple sentences and speech was more fluent. A "disconnection syndrome" was proposed to explain the language deficit. (van Dongen HR et al. Blind, deaf and mute after a status epilepticus caused by hyperpyrexia from shigellosis - a case report with a four-year follow-up. Neuropediatrics Dec 1993;24:343-345). (Respond: Dr HR van Dongen, Dept of Child Neurology, Sophia Children's Hospital, 40 Dr Molewaterplein, 3015 GD Rotterdam, The Netherlands).

> **COMMENT.** A reversible case of Kluver-Bucy syndrome in a 7-year-old child suffering from *Shigella flexneri* encephalopathy is reported from the Hebrew Univ of Jerusalem, Israel. (Guedalia JSB et al. J Child Neurol 1993;8:313-315). He was apathetic, his affect was dull, he did not recognize common objects or his relatives, he touched and placed objects in his mouth impulsively, and he exhibited an insatiable appetite and signs of bulimia. Hypermetamorphosis, a tendency to be distracted by minute visual stimuli, was questionable, and abnormal sexual behavior was absent. The patient showed 4 of the 6 classical signs of the K-B syndrome, a rare occurrence in children, and recovery was previously unreported.
>
> Shigellae are chiefly waterborne, and foods were incriminated in only 8 of 366 outbreaks in one report, the organism spread by fecal contamination and improper food handling. (Environmental Poisons in Our Food. PNB Publishers, 1993). Children under 10 years of age are at greatest risk, and a neurotoxin produced by *Shigella shiga* has been implicated as a possible convulsive agent. The incidence of febrile convulsions with shigellosis is as high as 45% in some reports whereas shigella-negative diarrheas caused convulsions in less than 2%. The incidence was independent of the species of Shigella, that included Flexner and Sonne, dysenteries not associated with neurotoxin formation. (Millichap JG. Febrile Convulsions. New York, Macmillan, 1968).

INFANTILE SPASMS AND BIOTINIDASE DEFICIENCY

Two patients who developed infantile spasms at 1 month of age and were found to have biotinidase deficiency are reported from the Hacettepe Children's Hospital, Ankara, Turkey. The parents were consanguineous. Corticotropin had been prescribed initially with partial seizure control. When evaluated at 3 months because of seizure exacerbation, the infants were lethargic and hypotonic, one had alopecia and seborrheic dermatitis and the other's scalp hair was sparse. Metabolic and lactic acidosis developed, and

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