

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 Kliver-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