

COMMENT. Causes of progressive myoclonus epilepsy include Unverricht-Lundborg disease (ULD), Lafora's disease, MERRF, Kufs' neuronal ceroid lipofuscinosis, and sialidoses. ULD is relatively common in Finland. Cochiu JI et al, at the Montreal General Hospital, have performed linkage analysis in 8 families, including 4 of neither Baltic nor Mediterranean origin. They report that ULD diagnosed in areas remote from the Baltic maps to the same locus on chromosome 21 as the Finnish form, and suggest that all ULD, regardless of geographic origin, represent mutations of the same gene (Ann Neurol Nov 1993;34:739-741). Lafora's disease, characterized by intraneuronal inclusions, is a form of myoclonus epilepsy not linked to EPM1 and chromosome 21. The recent advances in molecular genetics of neurological disease are reviewed in two current journals: Rosenberg RN et al. Arch Neurol Nov 1993;50:1123-1265, and Martin JB. Ann Neurol Dec 1993;34:757-773.

TRAUMATIC DISORDERS

SHAKEN BABY SYNDROME

Non-accidental head injury, with particular reference to whiplash shaking injury, is reviewed by paediatric neurologists at the Royal Hospital for Sick Children, Edinburgh. Of 30 children with non-accidental head injury followed by the authors, 17 were due to shaking and 13 to impact. Subdural hematoma was diagnosed in 16 (53%), subarachnoid hemorrhage in 3, and intracerebral hemorrhage in 4. Eleven (37%) developed post-hemorrhagic hydrocephalus. Velocity, the number of shakes per second, and duration are important components of degree of injury. The most common age for whiplash abuse is 5 months, when the head is large in relation to the body, and the neck muscles and head control are weak. Shaking injuries are rare after the second year. Vascular, midbrain, and white matter shearing injuries, cerebral edema, and retinal and subdural hemorrhages result. The clinical diagnosis and medico-legal aspects of non-accidental head injury are discussed in detail. (Brown JK, Minns RA. Non-accidental head injury, with particular reference to whiplash shaking injury and medico-legal aspects. Dev Med Child Neurol Oct 1993;35:849-869). (Respond: Dr JK Brown, Royal Hospital for Sick Children, Sciennes Road, Edinburgh EH9 1LF, Scotland).

COMMENT. Despite an increase in support services and media publicity, non-accidental head injury remains the leading cause of death or long-term disability among child abuse cases in Scotland and in the USA. The American Academy of Pediatrics Committee on Child Abuse and Neglect review the "Shaken baby syndrome: Inflicted cerebral trauma" in the current issue of Pediatrics Dec 1993;92:872-875. Subtle or mild trauma is particularly difficult to diagnose, often mistaken for viral illness, feeding problems, or infant colic.