

disorder is genetic with a recessive inherited pattern. A site on chromosome 5 has been identified but not localized exactly.

Except for a recent report of elevated pipecolic acid, other biochemical theories including glutamic acid decarboxylase deficiency have not been confirmed. (See Ped Neur Briefs 2000;14:27; 2000;14:52). One report of hyperprolinemia type II in a girl aged 20 months with convulsions and encephalopathy responded to IV and subsequent oral pyridoxine. The proline metabolite that accumulates in this disorder inactivates vitamin B6.

TRAUMATIC BRAIN DISORDERS

BRAIN DAMAGE FOLLOWING INFLICTED HEAD INJURY

Detailed neuropathological studies, including immunocytochemistry for microscopic damage, are reported in 53 cases of non-accidental head injury in children examined by 2 neuropathologists at the Universities of London and Sheffield, UK. Only 7 of the series had admissions of child abuse. Thirty seven were infants, with age at injury ranging from 20 days to 9 months, and 16 children with injury sustained at 13 months to 8 years. Skull fractures were present in 36% of cases, usually parietal or occipital, acute subdural bleeding in 72%, and retinal hemorrhages in 71%. None had extradural hematoma. Raised intracranial pressure secondary to brain swelling was the cause of death in 82% of cases. Microscopic exam revealed signs of severe hypoxic brain damage in 77%, vascular axonal damage in 40%, and diffuse traumatic axonal damage in only 6%. Localized axonal injury to the craniocervical junction of the spinal cord was present in 11 cases, all infants. Age-related patterns of damage were significant. *Infants of 2-3 months* presented with apnea and skull fracture, showed craniocervical axonal damage, a thin subdural hemorrhage, but no extracranial injury. Eight infants with no signs of impact were assumed to have "shaken baby syndrome." They had presented with collapse or respiratory arrest. The brain was swollen, 7 had a thin film of subdural hemorrhage, and 5 had bilateral retinal hemorrhages. The pathology in this "shaken-only" group was not different from the 29 infants with evidence of impact. *Children over 1 year* had a greater incidence of severe extracranial, especially abdominal, injuries, larger subdural hemorrhages, and adult patterns of hemispheric white matter damage. Diffuse axonal injury was an uncommon sequel of inflicted head injury in children. (Geddes JF, Hackshaw AK, Vowles GH, Nickols CD, Whitwell HL. Neuropathology of inflicted head injury in children. I. Patterns of brain damage. Brain July 2001;124:1290-1298). (Respond: Dr JF Geddes, Department of Histopathology and Morbid Anatomy, Royal London Hospital, Whitechapel, London E1 1BB, UK).

COMMENT. Patterns of brain damage in inflicted head injury in children are different from adults and also vary with the age of the child. Extradural hematomas are rare, subdural hematomas are thin films not requiring neurosurgical intervention and never massive, and subarachnoid bleeding is also rarely clinically significant. Traumatic axonal damage is mainly focal, involving the lower brainstem and cervical roots, and only occasionally diffuse. Whereas infants have craniocervical axonal damage as a result of cervical hyperextension/flexion stretch injury ("shaken-bay syndrome"), children have a greater incidence of severe extracranial, abdominal injury, and larger subdural hematomas with non-accidental inflicted head injury.

The authors have followed this paper with part II. Microscopic brain injury in infants (Geddes JF et al. Brain July 2001;124:1299-1306). The findings corroborate those outlined above.