

Ventricular shunt was performed in 14 infants, with resolution of symptoms in 7 (in 5 of 8 with grade I, 2 of 4 with grade II, and none of 2 with grade III symptoms). Of 10 with posterior fossa decompressions, symptoms resolved in only 2 (in 1 of 4 with grade I, one of 2 with grade II, and none of 4 with grade III symptoms). Within 6 months after symptoms began, one infant with grade II and 3 with grade III died. No deaths occurred with the grade I group. Infants with grade II or III symptoms have more extensive brain stem damage, such as hemorrhage, infarction and necrosis, and carry a poor prognosis whereas those with grade I symptoms often improve after neurosurgical procedures. (Charney EB et al. Management of Chiari II complications in infants with myelomeningocele. J. Pediatr 1987;111:364-71).

COMMENT: The grading of cases according to complications is useful in investigation, treatment and prognosis. In a previous study from the University of Toronto (Park TS et al. Neurosurgery 1983;13:147), decompression was recommended before rapid neurologic deterioration takes place, even if a functioning shunt is present. Of 45 infants with surgical decompression of the Chiari malformation, 28 survived and showed improved neurologic function and in 24 of these, recovery was complete. About 71% died of those patients who developed cardiorespiratory arrest, vocal cord paralysis, or arm weakness within 2 weeks before decompression, compared with 22% of those with more gradual neurologic deterioration.

BRAIN TUMORS

SUBARACHNOID HEMORRHAGE FROM BRAIN TUMORS

Six children with subarachnoid hemorrhage as the initial symptom of brain tumor are reported from the Depts of Neurosurgery, Univ. of Occupational and Environmental Health, Kitakyushu, and Kumamoto Univ Med Sch, Kumamoto. They represented a 3.6% of 167 new pediatric cases of brain tumor seen in 7-17 years at 2 centers in Japan. Two neonates presented with irritability, vomiting, cyanotic spells, and unilateral facial paresis. Four children, ages 4 to 15 years, developed sudden headache and vomiting with or without alteration of consciousness. The tumor locations were posterior fossa (2 medulloblastomas, one ependymoma, one hemangioma) and hypothalamus (one astrocytoma and one unverified). All were located close to the III or IV ventricles. The ultimate prognosis was poor. (Yokota A et al. Child's Nerv Syst 1987;3:65-69).

COMMENT: Medulloblastoma is more apt to bleed than other neuroectodermal tumors in pediatric patients. Compared to brain tumors in adults, those in children bleed more frequently and are more commonly located in the posterior fossa. Brain tumor should be considered as a possible etiology of subarachnoid hemorrhage in the neonate and child.

MUSCLE DISEASE

CONGENITAL MYOTONIC DYSTROPHY

Ten infants with congenital myotonic dystrophy admitted to the Dept

Pediatrics and Neonatal Medicine, Royal Postgraduate Medical School, Hammersmith Hospital, London, 1982-86, were investigated by ultrasonography or CT brain scans between 1 day and 2 months of age. The infants presented with generalized hypotonia, facial diplegia, and respiratory and feeding difficulties, and the diagnosis was confirmed by demonstrating maternal myotonia.

Cerebral ventricular dilation was demonstrated in 8 (80%) infants, and 3 were scanned on the first day of life. Neonatal asphyxia occurred in 7, associated with intraventricular hemorrhage (IVH) in 2. One had subarachnoid hemorrhage and one showed infarcts in the white matter. The pathogenesis of ventricular dilation in congenital myotonic dystrophy was probably IVH in 2, but a developmental anomaly during fetal life was the more likely explanation in the remainder. The authors note that mental retardation in 70% of cases can be related to the ventricular dilation which may be progressive and require surgical treatment. (Regev R, Dubovitz V et al. Cerebral ventricular dilation in congenital myotonic dystrophy. J Pediatr 1987;111:372-6).

COMMENT: It may be impolite to shake hands with a lady! But a handshake for a mother of a floppy baby with respiratory distress may be diagnostic of myotonia and is good clinical practice. Dr. Koh of Hope Hosp, Salford, England, asks the question "Do you shake hands with mothers of floppy babies?" as the title to his article on congenital myotonic dystrophy (Br Med J 1984;289:485).

BEHAVIOR AND LEARNING DISABILITIES

SERUM FATTY ACIDS AND HYPERACTIVITY

Serum essential fatty acids (EFA) levels were measured in 44 hyperactive children and 45 age-and-sex-matched controls at the Dept. of Pediatrics and Psychiatry and Behavioral Science, Univ. of Auckland, New Zealand. Docosahexaenoic, dihomogammolenic, and arachidonic acid levels were significantly lower in hyperactive children than controls. The hyperactive group of children had significantly lower birth weights than controls (3,058 and 3,410 g respectively; $p < 0.01$), a greater incidence of learning difficulties and dyslexia, but no increase in asthma, eczema, or other allergies. In a double-blind, placebo controlled, crossover study of evening primrose oil in 31 hyperactive children, effects on behavior were modest and equivocal. (Mitchell EA et al. Clinical characteristics and serum essential fatty acid levels in hyperactive children. Clin Pediatr 1987;26:406-411).

COMMENT: The search for dietary related causes and treatments for hyperactive behavior continues and now involves fats in addition to food allergies, additives, preservatives, sugar and megavitamins. In support of fats, a beneficial effect of the ketogenic diet on the behavior of the epileptic child often complements its anticonvulsant properties in my experience. The present paper did not confirm previous reports of a high prevalence of allergy among hyperactive children and tends to minimize the possible importance of food allergy as an etiologic factor.