

logical report of two cases with a contribution to pathogenesis and differential diagnosis. Clin Neuropath Sept/Oct 1990; 9:244-250).

COMMENT. Cerebral hemiatrophy with homolateral hemihyper-trophy of the skull and sinuses was described by Dyke CG, Davidoff L, and Masson C. (Arch Neurol and Psychiat 1933; 29:412) and Alpers B J, and Dear R B (J Nerve and Ment Dis 1939; 89:653) distinguished the primary and secondary groups of cases. The significance of convulsions in the pathogenesis of cerebral hemiatrophy is emphasized by many authors - the "hemiconvulsion hemiplegia - epilepsy syndrome." Permanent hemiparesis as a direct result of a febrile convulsion is a rare complication occurring in less than 0.2% (Millichap J. G. Febrile Convulsions, Macmillan, New York, 1968).

#### CHARGE SYNDROME

A 12 month old male infant with the CHARGE syndrome (Coloboma, Heart defect, Atresia of the choana, Retarded growth and development, Genital hypoplasia, Ear anomalies or deafness) is reported from the Departments of Pediatrics and Neurology, Loyola University, Chicago. The infant was admitted with repeated cyanotic episodes and generalized seizures. CT revealed agenesis of the cerebellar vermis and a large fourth ventricular cyst. Brain stem auditory evoked responses showed dysfunction of the distal VIII nerve (Menezes M, Coker S. B. CHARGE and Joubert's Syndromes, Are They a Single Disorder? Pediatr Neurol Nov/Dec 1990; 6:428-430).

COMMENT. This patient also had Joubert's syndrome (agenesis of the cerebellar vermis, respiratory irregularities, ocular colobomas). Chromosomes were normal. The authors believe that these two syndromes are a single disorder with an overlap of characteristic features.

Various associated abnormalities of brain development may be detected by studies of evoked potentials (Coupland S G, Sarnat H B Visual and auditory evoked potential correlates of cerebral malformations, Brain Dev 1990; 12:466-72). Auditory brain stem responses were abnormal in cases of holoprosencephaly, lissencephaly, pachygyria and generalized megalencephaly. Visual evoked potentials were normal in collosal agenesis, holoprosencephaly and colpocephaly; they were abnormal in septal optic dysplasia and optic nerve hypoplasia.

#### MUSCLE DISORDERS

##### MYOPATHY WITH COMPLEX I DEFICIENCY: RIBOFLAVIN CARNITINE THERAPY

A six year old boy with progressive myopathy and motor neuropathy associated with complex I deficiency is reported from the Institutes of