may interfere with closure. Meningomyelocele may be diagnosed in the fetus with 75 to 80% accuracy by measurements of serum alpha-fetoprotein levels, and ultrasonography can identify the extent and location of these defects. (Hobbins, J.C., Diagnosis and management of neural tube defects today, N Engl J Med, March 7 1991; 324:690-691).

The outcome of a neural tube defect may be improved by cesarean section before labor begins (Luthy, D A et al Cesarean section before the onset of labor and subsequent motor function in infants with meningomyelocele diagnosed antenatally, N Engl J Med March 7, 1991; 324:662-666). Infants who had been exposed to labor were 2.2 times more likely to have severe paralysis than those delivered by cesarean section without labor. The mean functional motor level was at the second lumbar vertebra in fetuses exposed to labor and at the fourth lumbar vertebra in those delivered before labor began. Cesarean section before onset of labor may result in better subsequent motor function and the ability to walk with minimal mechanical assistance.

CEREBRAL HEMIATROPHY: ETIOLOGY AND PATHOLOGY

Two types of cerebral hemiatrophy are reported from the Institute of Neuropathology at the University of Giessen, Arndtstrafse FRG. A case of "primary cerebral hemiatrophy" is described in a 12 year old boy whose birth was difficult and complicated by perinatal asphyxia. Seizures occurred immediately after birth and the psychomotor development was slow. He had spastic diplegia, asymmetrical, the right side predominating. He died at 12 years of age with cardiac arrest after status epilepticus. At autopsy the left hemisphere was reduced in size, the left lateral ventricle was enlarged and the white matter was decreased, especially on the left. The cerebral peduncles and pyramidal tracts were asymmetrical, and the neurons of the left hippocampus were replaced by glial tissue. Case two, an adult male, had a normal birth and early development and suffered from convulsions at two years of age which were followed by a right hemiparesis and homonymous hemianopsia. CT scan showed cerebral hemiatrophy and skull hypertrophy on the left side. The patient died at the age of 58 from intestinal hemorrhage with liver cirrhosis. The left cerebral hemisphere was atrophied and with crossed cerebellar and pyramidal tract atrophy. There was loss of cortical neurons in the atrophic left hemisphere and replacement by glial cells, a spongy state and abundant corpora amylacea. This was an example of "secondary cerebral hemiatrophy". The authors propose a classification of primary and secondary cerebral hemiatrophy. The primary forms are caused by vascular malformation, perinatal asphyxia and birth trauma and result in ulegyria, lamina necrosis and leukoencephalopathy. The secondary forms may be post-ictal with or without fever, cerebrovascular accident or leukoencephalitis. The pathological findings with the secondary form are sclerosing cortical atrophy and occasionally leukoencephalopathy (Vosskamper M, Schachenmayr W. Cerebral hemiatrophy: a clinical patho-

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 hemihypertrophy 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 (Menenzes 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