

COMMENT. Congenital hydrocephalus occurs in 0.5-1.8/1000 births. Subarachnoid hemorrhage at the time of birth has long been invoked as a cause of hydrocephalus (Paine RS. In *Pediatric Neurology*, Ed. Millichap JG, *Ped Clin N Amer* 1967;14:779), but the intrauterine development of posthemorrhagic hydrocephalus has been reported only recently. The authors encountered their 4 cases in a period of 2 years so that the incidence may be higher than the literature suggests.

CONGENITAL ARTHROGRYPOSIS AND MATERNAL MYASTHENIA

An infant with arthrogryposis multiplex and other malformations born to a mother who presented with myasthenia gravis immediately following cesarean section is reported from the Depts Neonatology and Obstetrics, Hasharon Hospital, Petah-Tiqva, Tel Aviv Univ Med Sch, Israel. The baby had hypotonia, absent suck, weak cry, and incomplete Moro. Multiple malformations included craniofacial dysmorphism, kyphoscoliosis, ventration of the diaphragm, and flexion contractures of the limbs. A Tensilon test at 7 days was negative and treatment with Mestinon for 3 weeks was without benefit, the infant dying at 5 weeks of age. Cytogenetic studies on infant and mother were normal. The authors suggest that the failure to recognize and treat the myasthenia gravis during pregnancy may be causally related to the infant's multiple malformations and fatal outcome. (Dulitzky F et al. An infant with multiple deformations born to a myasthenic mother. *Helv paediat Acta* Dec 1987;42:173-176).

COMMENT. Electromyography and nerve conduction studies, muscle biopsy and serum CPK may assist in determination of the site and nature of the pathology in cases of arthrogryposis. These tests were apparently not performed in the present case and the underlying cause of the hypotonia was not defined, except to rule out a transient neonatal form of myasthenia. A fetal form of spinal muscular atrophy, as described originally by Beever CE (*Brain* 1902;25:85) and later by Brandt S (*Acta paediat* 1947; 34:365), in association with arthrogryposis multiplex congenita, seems a more likely explanation for the fatal outcome than the maternal myasthenia.

PERINATAL ASPHYXIA

SERUM CPK AND HYPOXIC-ISCHEMIC ENCEPHALOPATHY (HIE)

The value of brain-typical creatine phosphokinase iso-enzyme (CPK-BB) determinations in the assessment of brain damage due to neonatal asphyxia in 33 full-term infants has been assessed in the Services of Neonatology, Child Neurology and Radiology, Clinica Infantil, Ciudad Sanitaria "La Paz", Madrid, Spain. Serum CPK activities measured at 4 hrs after birth were significantly higher in infants who died of HIE or developed neurological sequelae than in those asphyxiated infants who showed no neurological abnormalities during a 16 mo follow-up period or in a control group of 20 infants delivered normally. At 10 hrs after birth, the enzyme levels in brain damaged infants had decreased markedly and overlapped with values obtained in infants without sequelae at follow-up. Compared to the neurological exams, EEG's, and CT scans, obtained in the first or second week of life, the CPK assay was inferior as a predictor of neurological outcome. Normal CPK, CT and EEG's, and mild clinical encephalopathy were 96-100% predictive of a favorable outcome in asphyxiated full-term infants. (Fernandez F et al. Serum CPK-BB isoenzyme in the assessment of brain damage in asphyctic term infants. *Acta Paediatr Scand* Dec 1987;76:914-918).

COMMENT. In this study, an elevated serum CPK measured within 4 hrs after birth was a sensitive indicator of brain damage in asphyxiated term