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, eventration 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 Beevor 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, Cindad 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

infants but was of limited prognostic value in assessment of neurological outcome. These results contrast with those reported by Walsh P (J Pediat 1982;101:988), who found that serum CPK-BB activity measured in cord blood and at 6-12 hrs of life correlated with neurological outcome after severe asphyxia and compared favorably with CT scanning as a prognostic indicator. Normal CPK-BB activity was a predictor of good neurologic outcome in both studies (see Ped Neur Briefs 1987;1(3):17).

BRAINSTEM INJURY FROM PERINATAL ASPHYXIA

The clinical, radiological and neuropathological features of selective hypoxic-ischemic injury of the brainstem with relative sparing of cortex and subcortical white matter in an asphyxiated term infant are described in a case reported from the Division of Neurology and Depts of Pediatrics, Pathology and Radiology, British Columbia's Children's Hospital, Vancouver, Canada. The infant was pale, flaccid and without respiratory effort at birth and seizures occurred during the first hour. The Apgar score was one at 1, 5 and 10 min. The signs of brainstem dysfunction included abnormal horizontal eye movements, facial diplegia and ptosis, tongue fasciculations, and abnormal auditory evoked potentials. CT showed increased attenuation in the basal ganglia at 2 wks, and dilation of the third ventricle at 1 mo. Lateral ventricles and cortical sulci were normal, showing no atrophy.

The infant died of pneumonia at 4 mo of age. Neuropathological examination revealed scarring and pallor of the thalamus, basal ganglia and brainstem with neuronal loss and gliosis. (Roland EH et al. Selective brainstem injury in an asphyxiated newborn. Ann Neurol Jan 1988;23:89-92).

COMMENT. In animal studies, selective brainstem damage occurs after acute total asphyxia whereas the cerebral cortex and subcortical white matter are predominantly affected by prolonged partial asphyxia. In the human infant, the localization of hypoxic-ischemic encephalopathy is generally more diffuse (Volpe JJ Neurology of the Newborn 2nd ed, Philadelphia, Saunders, 1987) and selective brainstem injury is rare and frequently fatal.

CEREBRAL PALSY

A professor of obstetrics at the Univ of California at Davis School of Med, Sacramento, reviewing the relationship of obstetric care and management of asphyxia to the subsequent development of cerbral palsy (CP), refers to his own previously published study at Oxford University (Lancet 1984;2:827) and a similar study in progress at the Univ of Newcastle, England. Babies who were at risk for development of CP were compared with matched normal controls. The frequencies of substandard obstetric care were determined in the controls and in all cases of fetal death from asphyxia or trauma, those with severe asphyxia, convulsions in the first 48 hrs of life, and in children recognized to have CP at 18 mo of age.

Quality of care during labor proved to be less important than prenatal care. Substandard care during labor was not related to severe asphyxia, neonatal convulsions, or CP. A delay in the initiation of treatment for diagnosed asphyxia was not observed in CP cases, was uncommon in the control group (1.4%), but was frequent in cases of fetal death (20%), convulsions (7.9%) and severe birth asphyxia (5.4%). Substandard intrapartum care and especially the lack or failure to react appropriately to electronic fetal monitoring was causally related to neonatal seizures but not to CP.