diagnosis of early papilledema is often difficult, especially in small children, even for the experienced pediatric neurologist. Ophthalmologists may negate the neurologist's suspicions, but CT scan is nonetheless advisable if the clinical picture suggests a space-occupying lesion.

CONGENITAL MALFORMATIONS

NEURAL TUBE DEFECTS

In an overview of neural tube defects (NTD's), Dr RJ Lemire of the Dept Pediatrics, Univ of Washington and Children's Hospital, Seattle, WA, divides them into two major groups: (1) neurulation and (2) postneurulation defects.

Neurulation defects arising between the 17th and 30th day after fertilization are caused by nonclosure of the neural tube, leaving nervous tissue exposed, whereas postneurulation NTD's are covered by skin. Three general categories of neurulation defects are described: (1) craniorachischis (total dysraphism), (2) anencephaly, and (3) meningomyelocele. Environmental teratogenic factors implicated in neurulation defects include valproate sodium and nutritional and vitamin deficiencies (see Ped Neur Briefs 1987;1:15). Prenatal diagnosis is made by screening for maternal serum a-fetoprotein (AFP) levels during the 16th-18th week of pregnancy with follow-up ultrasound and amniocentesis when AFP is elevated. Elevated amniotic fluid acetylcholinesterase levels are confirmatory of open NTD and eliminate possible false-positive results of AFP tests. The population incidence of open NTD's is about 2/1000 births but the chance of recurrence is 1/20.

Postneurulation or closed NTD's arising after the 30th day of fetal life include hydrocephalus, encephalocele, and lumbosacral lesions. The causes of hydrocephalus and associated abnormalities are listed as follows: Arnold-Chiari malformation with meningomyelocele, tumors and cysts, aqueductal stenosis, achondroplasia, tuberous sclerosis, Dandy-Walker syndrome, chromosome trisomy 13 and 18 anomalies, prenatal infection, and aneurysm of the vein of Galen. Comprehensive lists of lumbosacral NTD's and encephalocele syndromes are provided. Early resection of caudal NTD's is advised when practical. (Lemire RJ. Neural tube defects. JAMA Jan 22/29 1988; 259:558-562).

COMMENT. As an encouraging postscript to this depressing subject, the author notes a declining incidence of NTD's in several areas of the world, including the U.S., related in part to prenatal diagnosis, genetic counselling and nutritional supplementation. Folate treatment before and at the time of conception prevent recurrence of spina bifida. Exposure to spermicide contraceptives is not a risk factor. (See Ped Neur Briefs Aug 1987; 1(3):15). A late occurring intrauterine cause of hydrocephalus is reported in the following paper.

CONGENITAL HYDROCEPHALUS

Intrauterine intraventricular hemorrhage occurring about 2 weeks or more prior to birth was the cause of congenital hydrocephalus in 4 newborn infants reported from the Abteilung Neonatologie, Universitats-Kinderklinik, Rumelinstrasse 23; D-7400 Tubingen, FR Germany. Multiple pregnancy was an associated risk factor in 2 cases and a hemorrhagic diathesis was present or suspected in 2. Intrauterine diagnosis of subependynal/intraventricular hemorrhage may be made by sonography of the fetal brain when indicated, especially in multiple pregnancy, hemorrhagic diathesis by history, fetal growth retardation, and signs of distress. Postnatally, cerebral ultrasound, CT and examination of the CSF for siderophages may be confirmatory. (Leidig E et al. Intrauterine development of posthemorrhagic hydrocephalus. Eur J Pediat Jan 1988; 147: 26-29).

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