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).