and 75% of those over 60 years old. Seizures developed in 84% of demented individuals with Down syndrome and 20% had Parkinsonian features. Hypothyroidism had been treated in 59% of the demented patients. CT scans showed brain tissue loss most pronounced in the temporal lobes. Neuropathological examination of 12 autopsied demented cases of Down syndrome showed gyral and central atrophy especially of the temporal lobes, and large numbers of plaques and tangles distributed in the same locations (i.e. hippocampus, amygdala, neocortex) as in the non-Down syndrome cases of Alzheimer dementia. (Lai F, Williams RS. A prospective study of Alzheimer disease in Down syndrome. Arch Neurol Aug 1989; 46:849-853).

COMMENT. The early age at onset of dementia in the Down syndrome population corresponds to the average age of onset (before age 52) in several large pedigrees of familial Alzheimer's disease. The gene for this form of autosomal dominant early onset Alzheimer disease has been mapped to the long arm of chromosome 21. An increased frequency of Down syndrome has been reported among relatives of early onset Alzheimer disease probands. The neuropathology and neurochemistry of Alzheimer's disease in aging individuals with Down syndrome and in the general population seem to be identical although the clinical expression of Alzheimer disease in Down syndrome shows some distinctive features, e.g. a high incidence of seizures.

HAW RIVER SYNDROME

A newly defined familial disorder of progressive dementia, ataxia, chorea, and seizures is described from the Department of Neurology School of Medicine, the University of North Carolina at Chapel Hill and the Department of Pathology, Duke University Medical Center, Durham, N.C. The first recorded member of the family was born in 1840 and lived at Haw River, N.C. In 22 patients examined the initial symptoms were ataxia of gait, intention tremor and choreiform movements that developed usually between 15 and 30 years of age. Recurrent generalized tonic-clonic seizures and progressive dementia developed later, and 11 of the 22 died after 15-25 years of illness. Neuropathological findings in two deceased family members were: neuronal loss of the dentate nucleus, microcalcification of the globus pallidus, neuroaxonal dystrophy of the nucleus gracilis, and demyelination of the centrum semiovale. (Farmer TW et al. Ataxia, chorea, seizures and dementia. Pathologic features of a newly defined familial disorder. Arch Neurol July 1989; 46:774-779).

COMMENT. The authors list in the differential diagnosis: Olivopontocerebellar atrophy, dentatorubropallidoluysian atropy, Ramsay Hunt syndrome, familial idiopathic calcification of the basal ganglia, neuroaxonal dystrophy, Hallervorden-Spatz disease, Huntington's disease, Wilson's disease, and Gerstmann-Straussler syndrome (cerebellar ataxia, dementia, amyloid plaques). Farmer's syndrome appears to have distinctive features.

Dementia as defined by the American Psychiatric Association (1980) is a deterioration in cognitive abilities that exceeds the decline

expected with normal aging and occurs in a state of clear consciousness. Classifications of the dementias have been based on etiology (degenerative, vascular, toxic metabolic, and infectious), pathology, and clinicopathological correlations. Cortical versus subcortical forms have been described and have been correlated with brain behavior relationships. In Alzheimer's disease corticopathology is prominent whereas in Parkinson's disease and Wilson's disease subcortical areas are the major sites of pathology. This dichotomy is probably an oversimplification. (See Chui HC. Arch Neurol July 1989; 46:806).

NUTRITION AND CNS DISORDERS

VITAMINS AND NEURAL TUBE DEFECTS

The use of vitamin supplements by women around the time of conception was examined and compared in those having babies with neural tube defects, those with still births or some other type of malformation, and in women who had normal babies. The study was performed at the National Institute of Child Health and Human Development, National Institutes of Health, Bethesda, Maryland; Northwestern University, Chicago; and the California Public Health Foundation, Berkley. The rate of periconceptional multivitamin use among mothers of infants with neural tube defects (15.8%) was not significantly different from the rate among mothers in either the abnormal or the normal control group (14.1% and 15.9%, respectively). There were no differences among the groups in the use of folate vitamin supplements. The authors conclude that the periconceptional use of multivitamins or folate-containing supplements did not decrease the risk of having an infant with a neural tube defect. (Mills JL et al. The absence of a relation between the periconceptional use of vitamins and neural-tube defects. N Engl J Med, August 17, 1989; 321:430-5).

COMMENT. Several studies have suggested that women who take multivitamins or supplements of folic acid around the time of conception may have a reduced risk of delivering an infant with a neural tube defect such as myelomeningocele or spina bifida. British studies have reported that folic acid in a dose of 4 mg/day or multivitamins can reduce the risk of recurrence in women who have already delivered an infant with such a defect. In a report published from the Atlanta Birth Defects Case Control Study, mothers of children with neural tube defects were significantly less likely to report vitamin use around the time of conception than were the mothers of infants with other malformations or normal control children. The results of the present study were strikingly different from those of the Atlanta Birth Defects Case Control Study in which 7% of mothers with affected babies and 50% of controls reported using multivitamin supplements at least three times a week in the periconceptional period. It is possible that the use of vitamins was not itself protective but was a marker for other health conscious behavior that prevented the malformations. Other explanations for the difference in the results might include the variation in the years studied and geographic differences. It should be noted that