syndrome.

Fetal valproate syndrome (FVS) and autism are reported in a 5 year-old boy exposed to valproic acid (VPA) in utero. (Williams PG, Hersh JH. Dev Med Child Neurol Sept 1997;39:632-634). The mother had taken VPA (500 mg 4xday) for 2 to 3 years before and through the 5th month of pregnancy. The infant's speech and language were delayed, and he communicated primarily by gestures. His head circumference was 55.5 cm (>95th centile), the forehead high and bossed, the nasal bridge flat, and the upper lip thin. Neurologic abnormalities included hypotonia, hyperreflexia, and ankle clonus. This is the second report of FVS manifested by autism.

LEARNING DISABILITY SUBTYPES IN NEUROFIBROMATOSIS 1

An analysis of neuropsychological data for 72 children, aged 6 to 18 years, with neurofibromatosis 1 (NF-1) and academic deficiencies is reported from the University of Texas MD Anderson Cancer Center, Houston, TX. Patients treated with cranial irradiation (6%) were excluded. Those with MRI areas of hyperintensity were not excluded. The IQs of the groups analyzed ranged from 78 to 107, and outlier groups (<5 patients) with above average IQs or the mentally deficient were excluded. Three groups of academic underachievers were identified: 1) neuropsychologically normal (39%); 2) a group with general academic deficiencies (47%); and 3) a group with visuospatial-constructional and fine motor coordination deficiencies, and without language deficits (14%). The low incidence of visuospatial deficiencies and absence of a group with pure language deficits were remarkable. (Brewer VR, Moore BD III, Hiscock M. Learning disability subtypes in children with neurofibromatosis. Jrnl of Learning Disabilities Sept/Oct 1997;30:521-533). (Respond: Bartlett D Moore III, Division of Pediatrics (Box 87), UTMD Anderson Cancer Center, 1515 Holcombe Blvd, Houston, TX 77030).

COMMENT. By using cluster analysis and exclusion of outlier groups with above average or retarded IQ scores, children with neurofibromatosis 1 who are academic underachievers may fall into one of three groups psychologically. Approximately 40% test normal, 50% have general learning disabilities, and 14% have visuospatial and motor coordination problems.

Correlations of cognitive impairments with MRI abnormalities and unidentified bright objects (UBOs) are of interest. A relation between the number or location of UBOs on MRI and cognitive deficits in children with NF-1 have been reported. The University of Texas study group (Moore et al, 1996) found location important, UBO hyperintensities in the thalamus correlated with a lowered IQ, whereas the Johns Hopkins group (Denckla et al, 1996) found the number of locations occupied by UBOs accounted for the lowering of IQ. (See Progress in Pediatric Neurology III, PNB Publishers, 1997; p291-294, p441-442, for articles and commentary on IQ and MRI findings in NF-1).

SEIZURE DISORDERS

GENETICS OF FAMILIAL INFANTILE CONVULSIONS

Four families from northwestern France with autosomal dominant benign infantile convulsions and paroxysmal choreoathetosis were studied genetically by linkage analysis at the Wellcome Trust Centre for Human Genetics, University of Oxford, UK, and at Genetic Clinics in Amiens, France. Linkage for the disease gene was found in the pericentromeric region of

human chromosome 16. (Szepetowski P, Rochette J, Berquin P, Piussan C, Lathrop GM, Monaco AP. Familial infantile convulsions and paroxysmal choreoathetosis: a new neurological syndrome linked to the pericentromeric region of human chromosome 16. Am J Hum Genet Oct 1997;61:889-898). (Reprints: Dr Anthony P Monaco, The Wellcome Trust Centre for Human Genetics, University of Oxford, Windmill Road, Headington, Oxford OX3 7BN, UK).

COMMENT. This study provides genetic evidence of an epileptic basis for paroxysmal choreoathetosis. Clinical features in common with epileptic seizures include paroxysmal attacks, tendency to remit, EEG epileptiform abnormalities, and response to antiepileptic drugs. A focus in the basal ganglia has been proposed for this syndrome. In differential diagnosis, choreoathetosis is a rare side effect of several AEDs. (Progress in Pediatric Neurology III, 1997;p157).

TEMPORAL LOBE EPILEPSY ETIOLOGICAL CLASSIFICATION

Clinical, EEG, and neuroimaging findings in a community-based cohort of 63 children with new-onset temporal lobe epilepsy (TLE) studied prospectively are reported from the Royal Children's Hospital, Melbourne, Australia. Three etiologically defined subgroups of TLE are proposed based on clinical history and MRI or CT findings: 1) *Cryptogenic,* with normal MRI and negative past history (54%); 2) *Hippocampal sclerosis,* or previous illness (29%); and 3) *Developmental,* malformation or tumor (16%). Febrile seizures had preceded onset of TLE in 13 children, and bacterial meningitis in 4. Neuroimaging revealed structural abnormalities of the temporal lobe in 24 (38%); hippocampal sclerosis in 13 (21%), tumor in 8 (13%). Focal temporal EEG abnormalities were recorded in 19 of 24 with lesions on MRI or CT and in 27 of 39 with normal neuroimaging. Developmental, behavioral, or learning problems occurred in 38% of the cohort. Behavior problems included hyperactivity (14), aggressiveness (13), and rage attacks (5). (Harvey AS, Berkovic SF, Wrennall JA, Hopkins IJ. Temporal lobe epilepsy in childhood: Clinical, EEG, and neuroimaging findings and syndrome classification in a cohort with new-onset seizures. Neurology October 1997;49:960-968). (Reprints: Dr A Simon Harvey, Department of Neurology, Royal Children's Hospital, Parkville, Victoria 3052, Australia).

COMMENT. This extensive prospective study has provided a classification of temporal lobe epilepsy in childhood of value in prognosis and management. Temporal lobe epilepsy in childhood is classified etiogically in three groups: 1) cryptogenic, 2) hippocampal sclerosis, and 3) developmental. Children with cryptogenic seizures have a relatively good prognosis, whereas those with hippocampal sclerosis or developmental lesions are at risk of resistant seizures and psychological disabilities. For previous reports from the Melbourne, Australia group, the Cleveland Clinic experience, and that of the University of California, Los Angeles, see Progress in Pediatric Neurology II, 1994;pp78-79. Vol III, 1997;pp54-55.

SPECT-EEG correlations in temporal lobe epilepsy are reported from Yonsei University Coll Med, Severance Hospital, Seoul, Korea. (Lee BI, Lee JD, Kim JY, et al. Neurology Oct 1997;49:981-991). Interictal SPECT correctly lateralized 8/9 patients with unitemporal epileptiform discharges and 5/10 with bitemporal EEG discharges. Ictal SPECT was highly concordant with ictal EEG, but correctly lateralized the lesion in only 11/19.