processing, and standard scales of intellectual functioning and reading. Serum levels of carbamazepine (CBZ) showed a significant negative correlation with changes in scores on the sum of five memory tests from before medication to 6 months and a year later. No significant correlations with memory scores were found for children treated with phenytoin (PHY). Only 10 of 126 serum level estimations of CBZ gave a value greater than 8.2 mcg/L. In the sodium valproate (SV) group significant positive correlations were found between serum levels and the sum of five memory tests at 1 month and at 6 months after starting treatment. The memory test clearly showed that there was impaired recent recall on CBZ apparent by 6 months of treatment and even more definite after a year. Reading scores were also lower in the CBZ compared with the PHY group after a year on drug treatment. Doses of CBZ were usually in the lower half of the accepted therapeutic range of 5-15 mcg/L (Forsythe I et al. Cognitive impairment in new cases of epilepsy randomly assigned to carbamazepine, phenytoin and sodium valproate. Dev Med Child Neurol June 1991; 33:524-534).

COMMENT. In this study involving children with epilepsy previously untreated, carbamazepine in moderate dosage adversely affected memory but sodium valproate and phenytoin did not. The tendency to favor carbamazepine in preference to phenytoin on the basis of reported cognitive deficits seems questionable and unfounded. A phenytoininduced improvement in auditory memory has been demonstrated in children with EEG dysrhythmias (Millichap J G et al. Auditory perceptual deficit correlated with EEG dysrhythmias. Response to diphenylhydantoin sodium. <u>Neurology</u> 1969; <u>19</u>:870-872).

METABOLIC AND DEGENERATIVE DISORDERS

CHILDHOOD DEGENERATIVE DISORDERS AND ADULT DEMENTIA

An approach to the evaluation of dementia in adults related to pediatric metabolic and degenerative diseases is reviewed from the Department of Neurology, University Medical Center, Maywood, IL. A list of 17 neurodegenerative disorders that ordinarily occur in childhood but may be present in adults includes adrenoleukodystrophy, metachromatic leukodystrophy, Krabbe's disease, Alexander's disease, Lafora's disease, Kufs' disease, mucopolysaccharidosis, Gaucher's type 1, Niemann-Pick disease, and gangliosidosis 1 and 2. Diagnosis is important for genetic counseling and some are treatable. A history of myoclonus suggests Kufs' disease, Lafora's disease, and mitrochondrial disorders. Easy tanning on sun exposure suggests Bone pain occurs with Gaucher's disease. adrenoleukodystrophy. Extrapyramidal symptoms suggest gangliosidosis. Muscle weakness is seen with gangliosidosis and mitochondrial disorders. Depressed deep tendon reflexes may occur with metachromatic leukodystrophy, adrenoleukodystrophy and gangliosidosis. Organomegaly is present in Gaucher's disease, Niemann-Pick disease and mucopolysaccharidosis. Macrocephaly can occur with Alexander's disease and metachromatic leukodystrophy. Special studies to be considered in young adults with idiopathic dementia include: 1) very long-chain fatty acids (adrenoleukodystrophy); 2) mucopolysaccharide screen; 3) arylsulfatase A (metachromatic leukodystrophy); 4) bone marrow for foam cells (Niemann-Pick disease); 5) copper and ceruloplasmin (Wilson's disease). (Coker S.B. Diagnosis of childhood neurogenerative disorders presenting as dementia in adults. <u>Neurology</u> June 1991; 41:794-798).

COMMENT. This review should be of value to the adult neurologist specializing in Alzheimer's disease and other causes of dementia. A knowledge of childhood neurodegenerative disorders would be important in the differential diagnosis of adult dementia.

ADRENOLEUKODYSTROPHY: CHILDHOOD AND ADULT FORMS COMPARED

Saturated, very long-chain fatty acids in erythrocyte membranes, blood plasma, and mononuclear cells were studied in 4 patients with childhoodadrenoleukodystrophy, adolescent 4 patients with adult adrenoleukodystrophy and 19 normal control subjects in the Department of Neurology, National Chikugo Hospital and the Neurological Institute, Kyusu University, Fukuoka, Japan. The ratios of C26:0 to C22:0 in patients were significantly higher than those of normal control subjects. Ratios of C26:0 to C22:0 in mononuclear cells were significantly higher in patients with childhood-adolescent adrenoleukodystrophy than in patients with adult adrenoleukodystrophy, whereas no significant difference in the ratios in erythrocyte membranes and blood plasma were noted. The results suggest that there is a correlation between phenotype and ratio of C26:0 to C22:0 within mononuclear cells in patients with adrenoleukodystrophy. (Antoku Y. et al. Adrenoleukodystrophy: a correlation between saturated very long-chain fatty acids in mononuclear cells and phenotype (Ann Neurol July 1991; 30:101-103).

COMMENT. Adrenoleukodystrophy is an X-linked recessive disorder characterized by adrenal insufficiency and demyelination of the CNS along with accumulation of saturated very long-chain fatty acids in tissues. Adrenoleukodystrophy has a wide phenotypical variation including childhood adolescent forms, adrenomyeloneuropathy with adult onset and adult rare forms. Variable phenotypes in a family kindred have been described. Marsh W.W. and Hurst D.L. reported a 20 month-old male who presented with a sudden onset of status epilepticus and cortical blindness; the patient died before his affected 5 year-old brother exhibited any features of ALD (Pediatr Neurol Jan-Feb 1991; 7:50-52).