The possibility of adrenomyeloneuropathy should be considered in any boy with Addison's disease. (Sadeghi-Nejad A, Senior B. N Engl J Med 1990; 322:13-16). These authors measured the plasma concentrations of very long chain saturated fatty acids in eight patients with adrenal insufficiency; five had elevated plasma hexacosanoic acid confirming the presence of adrenomyeloneuropathy. MRI showed brain involvement in all five patients. It was concluded that adrenomyeloneuropathy may present as Addison's disease in childhood.

In a further recent study from the Departments of Pediatric Endocrinology and Radiology, Hopital Saint Vincent de Paul, Paris, France, the MRI detected white matter lesions in two of seven patients with biochemically proven ALD but without neurologic manifestations. The ages at the time of MRI diagnosis were 14 and 21 years. (Auborg P et al. Neurology December 1989; 39:1619-1621). Six of the seven neurologically asymptomatic ALD patients in this study had adrenal insufficiency.

BIOGENIC AMINES IN RETT SYNDROME

The biogenic amines, dopamine, serotonin, and noradrenaline, and their metabolites, were measured in selected brain regions obtained at postmortem from four patients ages 12-30 years with Rett syndrome and are reported from the Departments of Pediatrics, Psychiatry and Neurochemistry, Goteborg University, Goteborg, Sweden. The cause of death was sudden and unexpected in one, severe pneumonitis and pulmonary abscess in one, in association with an operation for scoliosis at 12 years of age in one, and was unrecorded in one. Three of the patients had epilepsy; two were receiving carbamazepine and one sodium valproate at the time of death. Compared to determinations in two adults who had drowned in ice cold water and one killed in a traffic accident, the two older patients with Rett syndrome showed a 50% or greater reduction in biogenic amines in the substantia nigra whereas the youngest patient showed normal or nearly normal levels of biogenic amines in the substantia nigra. The levels were normal in the caudate nucleus, putamen and globus pallidus. The oldest patients had rigidity and dystonic posturing at the time of death whereas the younger 12 year old child was motor disabled secondary to weakness and wasting. The biogenic amine data reflect the clinical patterns of the patients and parallel the neuropathologic finding of reduced melanin content in the neurons of the substantia nigra. (Lekman A, Witt-Engerstrom I, Hagberg BA, Percy AK et al. Rett syndrome: Biogenic amines and metabolites in postmortem brain. Pediatr Neurol Nov-Dec 1989; 5:357-62). Dr. Percy is at the Department of Pediatrics, Baylor College of Medicine, Houston, TX.

COMMENT. Hagberg et al have previously reported a postmortem analysis of brain biogenic amines in an 11 year old Rett syndrome patient in whom the dopamine was markedly reduced in all regions of the brain except the cerebellum and parietal

cortex. (Ann Neuro 1983; 14:471). Motor dysfunction generally deteriorates steadily in Rett syndrome and parkinson-like features predominate during adolescence and early adulthood, suggesting a progressive involvement of the nigrostriatal system. Jellinger K, Percy AK et al. (Acta Neuropathologica 1988; 76:142) have described the autopsy findings in nine girls with Rett syndrome, ages 4-17 years. All brains were smaller than normal, lipofuscin was deposited in neuronal cytoplasm, melanin was absent in substantia nigra, and indications of dopaminergic nigrostriatal dysfunction were suggested. The intensive search for a biological marker for Rett syndrome continues.

RETT SYNDROME AND HELLER DEMENTIA

Six girls with Rett syndrome and two boys with Heller dementia are reported and contrasted with children with classic autism from the Department of Neuroscience and Pediatrics, University of North Dakota School of Medicine, Grand Forks, North Dakota, and the Kennedy Institute, Johns Hopkins Medical Institutions, Baltimore, MD. The study was performed in response to a report that Rett syndrome may be a form of Heller dementia with a predilection for girls (Millichap JG. Lancet Feb 21, 1987; 1:440). All eight children differed from those with classic autism in that they had normal prenatal and perinatal periods, followed by marked developmental regression, after which they acquired few or no skills. The boys with a diagnosis of Heller dementia differed from the girls with Rett syndrome in terms of estimated prevalence, age at onset, stereotypic breathing patterns, midline hand stereotypies, hand and gait apraxia, and speech development. The authors found no stereotyped movements in their two patients with Heller dementia although these have been described in other studies. The patients showed similarities in the normal prenatal and perinatal periods, behavioral, social and psychomotor regression, and epilepsy. The authors suggested that these children should be distinguished from those with classic autism and should be classified as "pervasive disintegrative disorder, Heller type" and "pervasive disintegrative disorder, Rett type". (Burd L, Fisher W, Kerbeshian, J. Pervasive disintegrative disorder: Are Rett syndrome and Heller dementia infantilis subtypes? Dev Med Child Neurol October 1989; 31:609-616).

COMMENT. In 1908, almost 60 years before the first description of Rett syndrome, Heller reported an infantile dementia with symptoms and a course similar in some respects to that of Rett syndrome. By 1930, Heller had collected 28 cases of dementia in young children who previously had been entirely normal in development. Without antecedent illness, a change in mood and behavior was noticed. The children became irritable, negativistic, and disobedient; they had outbursts of temper without provocation; they showed signs of anxiety; and a mental regression led to a complete loss of speech and deterioration within a few months. Motor restlessness and stereotyped