cells with interior signals in seizure foci. These X chromosome movements were limited to epileptic foci and were not simply the consequence of generalized seizure activity. (Borden J, Manuelidis L. Movement of the X chromosome in epilepsy. Science Dec 23, 1988;242:1687-1691).

COMMENT. Specifically altered nuclear patterns may become established and create the genetic memory for intractable seizures. These changes in chromosome arrangements may be caused by various lesions, including trauma, developmental abnormalities, and toxic factors. These studies provide a new approach to the mechanism of kindling, based on structural rearrangements of the X chromosome rather than functional alterations of the neuronal membrane and synapses.

METABOLIC AND DEGENERATIVE DISORDERS

DYSTONIA AND INFANTILE GLUTARIC ACIDEMIA

Glutaric acidemia, an autosomal recessively inherited disease caused by deficiency of glutaryl-CoA dehydrogenase, was manifested by acute dystonia in 3 infants reported from the Children's Hospital of Pittsburgh, Pennsylvania. Onset of symptoms was at 6, 9, and 21 months, the earlier history being entirely normal. Two infants developed dystonic posturing and hypertonia and one had choreoathetosis and hypotonia. Treatment with a low lysine diet, vitamin B and carnitine supplements, and baclofen resulted in some improvement in one infant, no change in one, and the third infant died at 13 months of age. Pathological findings included cerebral and cerebellar atrophy, shrinkage of the putamen, and white matter vacuolation. CT scans and MRI showed prominent sylvian fissures, progressive loss of cerebral volume, especially temporal lobes and caudate nuclei, and increased ventricular and subarachnoid spaces. Unusual features of the disease in these patients included the acute onset of symptoms, absence of metabolic acidosis, and late onset at 21 months in one. Analysis of glutaryl-CoA dehydrogenase in skin fibroblasts is the only definitive diagnostic test for glutaric acidemia. The authors stress that the enzyme should be measured in any infant with persistent dystonia and/or choreoathetosis and CT evidence of atrophic changes around the temporal lobes and sylvian fissures, even when urine organic acid analysis for excess glutaric acid is negative. (Bergman I et al. Acute profound dystonia in infants with glutaric acidemia. Pediatrics Feb 1989;83:228-234).

COMMENT. Glutaric acidemia, first described 14 years ago (Goodman SI et al. Biochem Med 1975;12:12), is now considered a relatively common metabolic disorder with an estimated frequency in the Swedish population about equal to phenylketonuria or 1/30,000 newborns (Kyllerman M, Steen G. Arch Fr Pediatr 1980;37:279).