

# PEDIATRIC NEUROLOGY BRIEFS

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## DEMYELINATING DISEASES

### FAMILIAL INCIDENCE OF MULTIPLE SCLEROSIS (MS)

Age-adjusted familial rates for MS were determined in children and siblings of patients studied at the MS Clinic and Dept of Genetics, Health Sciences Centre Hospital, Univ British Columbia, Vancouver BC, Canada. The risk for these relatives to develop MS was 3 to 5%, which was 30-35 times the 0.1% rate for the general population in this relatively "high-risk" area. For female index patients with MS, the proportion of children affected was 5/797 or 0.6% (all girls) but the age-adjusted risk was 2.6%, four times the crude rate. The proportion of daughters affected was 5/386 (1.3%) with an age-adjusted risk of 5%. (Sadovnick AD, Baird PA. The familial nature of multiple sclerosis: age-corrected empiric recurrence risks for children and siblings of patients. Neurology June 1988;38:990-991).

COMMENT. The onset of MS in childhood is unusual, but the 50-fold increase in risk for daughters of female patients with MS should alert neurologists to this diagnosis in young children with suggestive symptoms. The concordance rate for MS among monozygotic twins is 26% compared to 2% for dizygotic twins (Ebers GC et al. N Engl J Med 1986;315:1638).

### PELIZAEUS-MERZBACHER DISEASE (PMD)

The value of the MRI in the diagnosis of PMD in a 14-year-old Japanese boy is reported from the Depts of Pediatrics and Radiology, Tokyo Children's Rehabilitation Hospital, Tokyo 190-12, Japan. The child presented with rotatory nystagmus at 2 weeks after birth, titubation at 6 months, spasticity at 1 year, and a masklike facial expression at 3 years of age. He was markedly retarded at 14 years and examination revealed spasticity, intention tremor, joint contractures, nystagmus and normal fundi. Head circumference, nerve conduction studies, and lysosomal enzymes were normal. CT showed ventricular enlargement, cerebellar atrophy, but normal appearing white matter. MRI demonstrated diffuse

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