changes in white matter with sparing of scattered small areas consistent with a "tigroid pattern" of myelin preservation characteristic of PMD. (Shimomura C et al. Magnetic resonance imaging in Pelizaeus-Merzbacher disease. Pediatr Neurol 1988;4:124-5).

COMMENT. Described by Pelizaeus in 1885 and by Merzbacher in 1910, this heredofamilial disease transmitted as an X-linked recessive character and occurring chiefly in males is a slowly progressive leukodystrophy with a long course, patients not infrequently surviving into middle age. When the family history is negative, as in the above case, confirmation of the diagnosis during life is difficult but may be facilitated by the MRI findings and may permit appropriate genetic counseling.

CONGENITAL AMYELINATING NEUROPATHY

A case of severe neurogenic arthrogryposis multiplex congenita caused by absence of peripheral nerve myelin in an infant who died at age 31 days of aspiration pneumonia is described from the Dept of Neurology, Neuromuscular Division, Johns Hopkins University School of Medicine, Baltimore, MD. The infant, delivered by Cesarean section, had Apgar scores of one at 1 and 5 minutes, and examination revealed multiple fixed joints, small chin and triangular face, bilateral extraocular and facial pareses, diffuse hypotonia, muscle atrophy, and areflexia. Absence of myelin in the peripheral nerves at autopsy reflected an arrest in the differentiation or maturation of Schwann cells at the stages of elongation and longitudinal growth of the mesaxon. The authors refer to serial ultrasonography to assess fetal movement after the 18th week, the time of onset of peripheral nerve myelination, as an antenatal diagnostic technique in such families, and as suggested by Miskin M et al. (Charnas L et al. Congenital absence of peripheral myelin: Abnormal Schwann cell development causes lethal arthrogryposis multiplex congenita. Neurology June 1988;38:966-974).

COMMENT. The most common cause of arthrogryposis multiplex congenita is probably an amyoplasia due to anterior horn cell maldevelopment and associated muscle atrophy. Other causes include anterior horn cell degeneration, congenital myopathies, mechanical interference with fetal movement, and peripheral neuropathy. Paralysis of fetal movement is the common link in all forms of the disorder. For a comprehensive account of arthrogryposis, the reader should refer to Clinical Orthopedics April 1985;194, an issue devoted entirely to the topic. A long-term follow-up study showed that 17 of 34 patients examined at 16 years of age or older were able to walk independently and 9 others walked with the aid of crutches or braces. The prognosis is obviously hopeful in a majority of cases with nonprogressive underlying causes, and appropriate orthopedic procedures can achieve correction and relative independence at an early age.

SEIZURE DISORDERS

POST-TRAUMATIC SEIZURES

Factors influencing the occurrence of post-traumatic seizures in 92 of 937 children with head injuries (9.8%) were studied in the Division of

Neurosurgery, Children's Memorial Hospital, Chicago, Illinois. Seizures were generalized in 64% and focal in 28%. They occurred within 24 hours after injury in 95% and within 7 days in 98%. The injury was caused by a fall in 60%. Children with a severe head injury (Glasgow Coma Scale $\leq$ 8) had a 7 times higher incidence of seizures than those with minor trauma. Those with CT evidence of diffuse cerebral edema or subdural hematoma had the highest incidence of seizures. Prophylactic use of anticonvulsants was recommended in children with diffuse cerebral edema, subdural hematoma, open depressed skull fracture, or severe head injuries. (Hahn YS et al. Factors influencing post-traumatic seizures in children. Neurosurgery May 1988;22:864-867).

COMMENT. Unfortunately, the duration of prophylactic anticonvulsant therapy was not addressed in this report, although the follow-up period was 7 months to 6 years. In a previous study at the Mayo Clinic involving 2747 patients of all ages with head injury, early seizures occurred in 2.1%. The risks of post-traumatic seizures after severe injury were 7.1% within 1 year and 11.5% in 5 years, after moderate injury 0.7% and 1.6%, and after mild injury 0.1% and 0.6%. Children were at a greater risk for early seizures after severe trauma than adults, but late seizures in children were less frequent and had no relation to the occurrence of early seizures. Mild head trauma in both children and adults did not cause epilepsy. (Annegers JF et al. Neurology 1980;30:683).

ABSTINENCE-ASSOCIATED NEONATAL SEIZURES (AANS)

The neurodevelopment of 14 infants with AANS was assessed during the first year of life in the Division of Neonatology (Dr Kandall), Beth Israel Medical Center, First Ave at 16th St, New York, NY. Bayley developmental scores remained normal and most early EEG and neurological abnormalities, including hypertonia, hyperreflexia, tremors and irritability, became normal during follow-up. Seizures, mainly myoclonic, were controlled initially with phenobarbital I.V. and then oral phenobarb or paregoric. Medications were gradually discontinued if EEG's reverted to normal. Of 9 original abnormal EEG's, 4 were normal by 8 weeks of age and only one remained abnormal at 6 months. Clinical improvement paralleled EEG improvement. Prognosis for AANS was good and different from that of neonatal seizures due to other causes. (Doberczak TM et al. One-year follow-up of infants with abstinence-associated seizures. Arch Neurol June 1988;45:649-653).

COMMENT. Infants born to methadone and heroin-dependent mothers have a reported risk of 20% and 4%, respectively, of developing seizures. Fortunately, these neonatal seizures appear to be transient and unassociated with persistent neurological deficits, whereas infants with neonatal seizures from other causes have a mortality rate of 35% and two-thirds of survivors suffer from cerebral palsy, epilepsy or retardation. (Holden KR et al. Pediatrics 1982;70:165).

EPILEPSY IN AUTISM

Epilepsy occurred in 27% (14/52) of children with autism under 10 years of age in a population-based study in the Dept of Child and Adolescent Psychiatry and Pediatrics, University of Goteborg, Sweden.