7. Visual evoked potentials (VEP) were abnormal in 7 of 8 patients; BAEP in 4 of 8: and SEP in 4 of 8. The importance of paraclinical examinations in diagnosis is emphasized. (Guilhoto LM de FF, Diament A et al. Pediatric multiple sclerosis report of 14 cases. Brain Dev Jan/Feb 1995;17:9-12). (Respond: Dr Laura Maria de Figueiredo Ferreira Guilhoto, Departamento de Neurologia do Hospital das Clinicas da Faculdade de Medicina da Universidade de Sao Paulo, Av Dr Eneas Carvalho Aguiar S/N, CEP 05403-900 Sao Paulo, SP, Brazil).

COMMENT. The 14 cases were seen in a period of 12 years. Four presented before 5 years of age, and the youngest was 2 years.

According to a report from the George-August University, Gottingen, Germany, published in the review International MS Journal (Hanefeld FA. Int. MSJ 1995;1:90-97), 24 cases with MS onset before age 5 years have been published since 1969. In 20 of 39 new patients studied over a 5-year period in Gottingen, the onset was before 10 years. The onset or relapse was preceded by a nonspecific infection, usually an URI, in >50%. Of 8 presenting with optic neuritis, 4 developed MS within 2 years. CSF maximal cell count was 900/ml, and protein was increased >100 mg%. Oligoclonal bands were absent in one third. VEPs were more frequently abnormal than BAEP and SEP. MRI sometimes showed new lesions without accompanying symptoms or relapse, and remissions were not always reflected in less MRI changes. Patients with juvenile onset (>10 years) followed a more severe, frequently relapsing, course than those with onset before puberty.

MYELIN DEVELOPMENT IN SIDS: MRI FINDINGS

The MRI brain scans of 28 SIDS infants were compared with 14 controls at the Neuropathology Unit, University of Sydney, and the Department of Radiology, Royal Prince Alfred Hospital, New South Wales, Australia. The amount of myelin assessed by densitometer in 21 of 26 sites showed no changes in 15 sites, and a higher rate of myelination in 6 sites, but only in infants older than 8 months. No focal white matter abnormalities were detected. (Lamont P et al. Myelin in SIDS: Assessment of development and damage using MRI. Pediatrics March 1995;95:409-413). (Reprints: Dr Roger Pamphlett, Department of Pathology, University of Sydney, New South Wales 2006, Australia).

COMMENT. This MRI investigation failed to confirm the histopathological evidence of delayed myelination in SIDS victims reported from the University of Toronto (Becker LE. Neural maturational delay as a link in the chain of events leading to SIDS. Can J Neurol Sci Nov 1990;17:361). See Progress in Pediatric Neurology I, 1991, pp309-310, for a review of mechanisms of SIDS. It was concluded that a central type of respiratory failure or cardiac dysrythmia was involved. A delayed development of the vagus nerve, similar to the finding in an infant with Ondine's curse, was described in the Canadian study.

The Steering Committee of Collaborative Home Infant Monitoring Evaluation reports on a multi-center study aimed at correlating events in infants at increased risk for SIDS, including siblings of prior SIDS victims. (Hunt CE. Sudden infant death syndrome and subsequent siblings. Pediatrics March 1995;95:430-432). It concluded that siblings are at increased risk for SIDS, and monitoring is cost-effective in sibs of prior SIDS infants.

INTERLEUKIN-6 CSF LEVELS were increased in 20 infants dying of SIDS in a study reported from the Institute of Forensic Medicine, National Hospital, Oslo, Norway. (Vege A et al. Acta Paediatr Feb 1995;84:193-6). The authors suggest that immune activation plays a role in SIDS, and cytokines in the CNS may cause respiratory depression in vulnerable infants.

An increased postneonatal mortality in lower social groups was explained by an association with SIDS in a study from the Department of Epidemiology, National Institute of Public Health, Oslo, Norway. (Arntzen A et al. Acta Paediatr Feb 1995;84:188-92).

A series of articles and an editorial in a recent issue of JAMA address the roles of sleeping position and passive smoking and tobacco exposure through breast milk in the etiology of SIDS. A major factor relating to a decline in SIDS in Tasmania was a reduction in the prevalence of prone sleeping position of infants. (Dwyer T et al. JAMA March 8, 1995;273:783-789). In contrast, routine prone sleeping position was not associated with an increased risk of SIDS in a Southern California study population. (Klonoff-Cohen HS, Edelstein SH. JAMA 1995;273:790-794). Passive smoking in the same room as infants increased the risk for SIDS in a study at the University of California, San Diego. (Klonoff-Cohen HS, et al. JAMA 1995;273:795-798). An editorial by Willinger M (JAMA 1995;273:818-819) advises that caregivers should follow AAP recommendations, and parents should be counselled that back or side sleep position is one measure to protect their infant from SIDS, but it is not fool-proof.

A CASE OF ACQUIRED "PSEUDO" HYPERTROPHIC NEUROPATHY

A 9-year-old boy with chronic progressive motor-sensory neuropathy beginning in early infancy and reversed by corticosteroid therapy is reported from the Institute of Neurological Diseases, Hirosaki University School of Medicine, Japan. The parents had noticed an awkward gait and frequent falling after learning to walk at 15 months of age. He was in a wheel chair at examination, and he complained of hand numbness. Limb muscles were severely weakened and atrophied, and intrinisic hand muscles totally paralysed. Pes cavus was bilateral. Tendon reflexes were absent. Nerves at elbows and knees and behind the ears were thickened and enlarged. CSF protein was 68 mg/dl. Biopsy of the sural nerve showed edematous swelling, and loss of myelinated fibers, but only occasional onion bulbs. One week after IV methylpredisolone (25 mg/kg/day) for 3 days, followed by oral prednisolone (2 mg/kg/day), numbness in the hands decreased, and sensation and muscle strength improved. Within four weeks, he was walking alone, and posterior auricular nerves were no longer visible. Comparison of EMG and NCS before and after steroids showed that the extremely slow conduction velocities of 2 m/s had increased to 7 to 16 m/s. (Baba M et al. "Pseudo" hypertrophic neuropathy of childhood. J Neurol Neurosurg Psychiatry Feb 1995;58:236-237). (Respond: Dr Masayuki Baba, Department-of Neurology, Institute of Neurological Diseases, Hirosaki University School of Medicine, Zaifu-cho 5, Hirosaki 036, Japan).

COMMENT. Steroid responsive neuropathy in childhood (Byers and Taft. Pediatrics 1957;20:517) was cited as the first reference to this disorder.