

UK. Only one child had symptoms of AAI over 5 years. A 9 year old developed acute torticollis after ENT surgery; radiographs showed a rotatory dislocation and a AA gap of 7 mm (>4 mm is abnormal). Of 7 with AAI at initial exam in 1986, 5 were abnormal in 1991, but none had symptoms. No additional cases of AAI were found at 5 years. In fact, a significant reduction of the mean AA gap from 2.95 mm to 2.45 mm occurred over 5 years. (A 1 mm difference in AA gap can occur between 2 of 3 radiographs taken 10 minutes apart). Radiographs were taken with maximal flexion of the upper neck. (Morton RE et al. Atlantoaxial instability in Down's syndrome: a five year follow up study. Arch Dis Child Feb 1995;72:115-119). (Respond: Dr Richard Morton, Ronnie MacKeith Child Development Centre, Derbyshire Royal Infirmary, London Road, Derby DE1 2QY, UK).

COMMENT. Chapman S, of the Radiology Department, Children's Hospital, Birmingham, cautions that 1) the use of a single flexion view is diagnostic of subluxation - not AA instability; 2) the apparent lessening of the gap with age may be difficult to distinguish from observed variations in repeat measurements; 3) the spine radiograph is not a good clinical predictor in Down's syndrome and, in the presence of symptoms or neurological signs, should probably be replaced by MRI or CT; and 4) films in flexion and extension should be obtained before anesthesia or before engaging in high risk sports. (Arch Dis Child 1995;72:118-119).

To avoid possible exacerbation of AAI and symptoms following recurrent trauma and hyperflexion of the neck, the authors restricted sports activities of the seven children who initially were found to have AAI on radiographic examination. Diving, trampolining, gymnastics, judo, and rugby were forbidden, and running, jumping, and swimming were encouraged. None developed symptoms. However, other reports suggest that restriction of activities is unnecessary, and radiographs prior to sporting activities are unjustified. The clinical examination and history of symptoms appear to be more important than radiographic screening tests. The management of this complication of Down's syndrome remains controversial. Parents should be made aware of potential cervical spine problems in a child with Down's syndrome.

NON-TRAUMATIC ATLANTO-AXIAL SUBLUXATION is reported from Mater Private Hospital, Dublin, Ireland, in an 8-year-old child with a previous history of neonatal retropharyngeal abscess. (O'Neill P. Child's Nerv Syst 1994;10:396-398). Limitation of neck movement was followed by cord compression and progressive quadriplegia in a period of 6 months. Function improved after anterior decompression and occipito-cervical fusion.

## DEMYELINATING DISORDERS

### CHARACTERISTICS OF CHILDHOOD MULTIPLE SCLEROSIS

The clinical manifestations of multiple sclerosis (MS) in 14 children are reported from the Universidade de Sao Paulo, Brazil. Age at onset ranged from 2 to 15 years. Initial symptoms varied from minor motor impairment, visual disturbances, bladder dysfunction and paresthesias, to a diffuse encephalopathy, with impaired consciousness. All had a relapsing-remitting course, and one died 6 months after onset with disseminated demyelinating lesions. CT showed demyelination in 6 of 9 patients. MRI showed white matter lesions in the brainstem or cerebral hemispheres in 5 of 6 patients. CSF pleocytosis occurred in 8 of 23 attacks, and g-globulin levels were increased in

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, Diamant 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 1, 1991, pp309-310, for a review of mechanisms of SIDS. It was concluded that a central type of respiratory failure or cardiac dysrhythmia 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.