BRAIN ACTIVITY DURING READING

PET was used to study the functional anatomy of reading in the intact brain of subjects examined at the Hammersmith Hospital, London, and other centers. The type of reading task and the exposure duration of the word stimuli were variables that influenced the patterns of brain activity. Three tasks were applied as follows: reading aloud, reading silently, and distinguishing words and pseudowords in a lexical decision task. Reading aloud and reading silently produced activity in the left posterior temporal lobe. Lexical decision involved the left inferior and middle frontal cortices and the supplementary motor area. Brain activity was greater for short exposure durations than for long durations. (Price CJ et al. Brain activity during reading. The effects of exposure duration and task. Brain Dec 1994;117:1255-1269). (Respond: Dr Cathy Price, MRC Cyclotron Unit, Hammersmith Hospital, Ducane Road, London W12 OHS, UK).

> COMMENT. Small variations in experimental design may influence brain activity as measured by PET, and the association of specific reading tasks with discrete anatomical areas must be interpreted with caution. The authors stress that the aim of their study was to determine reasons for inconsistencies in previous reports.

BEHAVIOR DISORDERS AND NOONAN'S SYNDROME

The behavioral phenotype and psychiatric symptoms of 21 children with Noonan's syndrome are reported from the Departments of Child and Adolescent Psychiatry, and Clinical Genetics, Royal Manchester Children's Hospital, UK. Ages ranged from 2 to 16 years (median, 9 years). Non-behavioral abnormalities included pregnancy complications (52%), short stature (71%), visual problems (57%), and language impairment (43%). Behavioral abnormalities in >50% of the group included clumsiness (71%), faddy eating habits (67%), fidgeting (62%), stubbornness (62%), echolalia (52%), euphoria (52%), and irritability (52%). On the Child Behavior Checklist for parent reported psychiatric abnormalities, only social relationships and attention span were disturbed, and these are non-specific. A behavioral profile for Noonan's syndrome is suggested by these findings but compared to sibling controls, the abnormalities were not statistically significant. (Wood A et al. Behavioral aspects and psychiatric findings in Noonan's syndrome. Arch Dis Child Feb 1995;72:153-155). (Respond: Dr A Wood, Department of Child and Adolescent Psychiatry, Royal Manchester Children's Hospital, Hospital Road, Pendlebury, Manchester M27 4HA, UK).

> COMMENT. Noonan's syndrome is a genetic disorder characterized by pulmonary stenosis, facial abnormalities, short stature, and learning difficulties. Few studies have emphasized behavioral abnormalities and a behavioral phenotype has not been delineated. The above behavioral characteristics may be helpful in diagnosis, counselling, and treatment. The association of Noonan's syndrome and neurofibromatosis has been described. (Listernick, Charrow, 1990; see Progress in Ped Neur II, 1994, p377).

ATLANTOAXIAL INSTABILITY IN DOWN'S SYNDROME

The radiographs and clinical evaluations of 90 children with Down's syndrome were reassessed after an interval of 5 years in a study of atlantoaxial instability (AAI) at the Derbyshire Children's Hospital and Infirmary, Derby,

UK. Only one child had symptoms of AAI over 5 years. A 9 year old developed acute torticollis after ENT surgery; radiographs showed a rotatary 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 quadriparesis in a period of 6 months. Function improved after anterior decompression and occipito-cervical fusion.

DEMYELINATING DISORDERS

CHARACTERISTICS OF CHILDHOOD MULTIPLE SCLEROSIS

The clinical manfestations 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