PEDIATRIC NEUROLOGY BRIEFS

A MONTHLY JOURNAL REVIEW

J. GORDON MILLICHAP, M.D., F.R.C.P., EDITOR

Vol. 3, No. 8 August 1989

MENTAL RETARDATION AND DEMENTIAS

WILLIAMS AND DOWN SYNDROMES

The neurological features of the Williams (WS) and Down (DS) syndromes were compared as part of a large multidisciplinary research center study and reported from the Departments of Neurosciences and Pediatrics, University of California School of Medicine, San Diego, CA. Eight patients with Williams syndrome and six with Down syndrome were matched for age (mean ages 16.7 and 15.8 years respectively) and WISC-R or WAIS scores revealed no significant differences between the two groups (WS: 53.8 ± 7.3; DS: 52.5 ± 8.8). DS patients demonstrated nonspecific features of global developmental delay but functioned fairly well for their developmental ages while those with WS demonstrated impaired oramotor skills, cerebellar dysfunction, difficulty with drawing, and higher verbal abilities than expected. WS patients also were small for gestational age and were more likely to have had early feeding problems and failure to thrive. One-half of the WS patients had epilepsy. The authors consider that neurologic distinctions between these two groups may reflect an underlying metabolic defect in Williams syndrome. (Trauner, DA et al. Neurologic featurse of Williams and Down syndromes. Pediatr Neurol May/June 1989; 5:166-8).

> COMMENT. Williams syndrome is a disorder of unknown etiology characterized by distinctive elflike facial features, mental retardation, cardiac defects and infantile hypercalcemia. A dissociation between language and cognitive skills described in patients with this disorder suggests a specific neuropsychologic profile. Seizures as a frequent manifestation of WS have not been reported previously.

ATLANTOAXIAL INSTABILITY IN DOWN SYNDROME

Results of an investigation of 130 children with Down syndrome screened for atlantoaxial instability are reported from Our Lady's

PEDIATRIC NEUROLOGY BRIEFS (ISSN 1043-3155) ©1989 covers selected articles from the world literature and is published monthly. Subscription requests ($28 US or £15 UK annually; add $5 (£3) for airmail outside North America) may be sent to: Pediatric Neurology Briefs - J. Gordon Millichap, M.D., F.R.C.P. - Editor, P.O. Box 11931, Chicago, IL 60611, USA, or P.O. Box 1605, London W8 7JD, UK. The Editor is Professor of Neurology and Pediatrics at Northwestern University Medical School, Chicago and is presently at SIU School of Medicine, Springfield, Illinois, USA.