PEDIATRIC NEUROLOGY BRIEFS
A MONTHLY JOURNAL REVIEW

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

Vol. 13, No. 9 September 1999

NEUROCUTANEOUS SYNDROMES

EYE MOVEMENT DISORDERS IN ATAXIA TELANGIECTASIA

Eye movements were examined clinically in 56 patients (age range, 2-25 years; mean, 10.7 years) with ataxia telangiectasia (A-T) at Johns Hopkins University, Baltimore, MD. Electrooculographic recordings of eye movements were obtained in 33 patients. Deficits occurred in eye movement systems that stabilize images on the retina (pursuit, gaze holding, convergence, vestibular and optokinetic slow phases, and cancellation of vestibular slow phases), and in systems that maintain fixation and shift gaze, characterized by abnormal reflexive and voluntary saccades, head movements associated with gaze shifts, ocular motor apraxia, impaired fixation, and a reduction in vestibular and optokinetic quick phases. Clinical oculomotor abnormalities increased with age and were more prevalent in patients with severe neurologic abnormalities, whereas electrooculographic signs were not age or neurologic sign related. Deficits in image stabilization are associated with dysfunction in the cerebellar flocculus and ventral paraflocculus, and fixation deficits are probably correlated with dysfunction in the cerebellar vermis or the basal ganglia which affects the superior colliculus. (Lewis RF, Lederman HM, Crawford TO. Ocular motor abnormalities in ataxia telangiectasia. Ann Neurol September 1999;46:287-295). (Respond: Dr Lewis, Department of Neurology, Harvard Medical School, 243 Charles Street, Boston, MA 02114).

COMMENT. Atrophy of the cerebellum, especially the vermis, is the most prominent structural abnormality in pathological and imaging studies of ataxia telangiectasia. Eye movement abnormalities involving stabilization of retinal images are localized in the cerebellum, whereas impairments of fixation and shifts in gaze are more likely correlated with dysfunction in the cerebellar vermis or basal ganglia.

CEREBRAL BLOOD FLOW IN STURGE-WEBER SYNDROME

Regional cerebral blood flow during seizure activity, measured by transcranial Doppler sonography, and SPECT were studied in three infants with

PEDIATRIC NEUROLOGY BRIEFS (ISSN 1043-3155) © 1999 covers selected articles from the world literature and is published monthly. Send subscription requests ($58 US; $60 Canada; $68 airmail outside N America) to **Pediatric Neurology Briefs - J. Gordon Millichap, M.D., F.R.C.P.-Editor,** P.O. Box 11391, Chicago, Illinois, 60611, USA.
The editor is Pediatric Neurologist at Children's Memorial Hospital and Northwestern University Medical School, Chicago, Illinois.
PNB is a continuing education service designed to expedite and facilitate review of current scientific information for physicians and other health professionals.