CEREBELLAR ATROPHY IN LEUKEMIA SURVIVORS

MRI changes in the cerebellum and cognitive function of 13 survivors of childhood acute lymphoblastic leukemia (ALL) treated with cranial radiation of 24 Gy and intrathecal methotrexate were studied at the University of New Mexico, Albuquerque, Manitoba Cancer Foundation, Winnipeg, and University of Alberta, Edmonton. Age at diagnosis was a mean of 3 yrs 3 mos (range, 2 yrs - 4 yrs 10 mos), and age at testing was 11 yrs 8 mos (9 - 14 yrs). Hypoplasia of the cerebellar vermis and cognitive deficits involving visual-spatial-motor coordination and memory were observed in patients compared to controls. (Ciesielski KT et al. Hypoplasia of the cerebellar vermis and cognitive deficits in survivors of childhood leukemia. Arch Neurol Oct 1994;51:985-993). (Reprints: Dr Ciesielski, Department of Psychology, University of New Mexico, Logan Hall, Albuquerque, NM 87131).

COMMENT. The neurotoxic effects of prophylactic cranial irradiation in children with acute lymphoblastic leukemia are well documented. As in this study, deficits in visual-motor skills and right brain involvement have been more pronounced than language and verbal deficits - evidence of left brain sparing. A causal relationship between the structural cerebellar abnormalities and the neuropsychological dysfunction is unproven. Children treated before 5 years of age are more susceptible to post-irradiation cognitive deficits than those diagnosed at or later than 5 years. Children in families of higher educational status who receive more attention and academic stimulation have a lower incidence of cognitive deficit than those with sensory-motor learning deprivation.

The dose of cranial irradiation, 2400 rads, used in this study is highly toxic in young patients. In the 1980s, most leukemia protocols limited the prophylactic dose to 1800 rads for use only in high risk patients. Reliance on intrathecal methotrexate chemotherapy for the majority of ALL patients is not known to result in significant intellectual or coordination deficits. (Personal communication: Dr David O. Walterhouse, Oncology Service, Children's Memorial Hospital, Chicago).

MOVEMENT DISORDERS

HEREDITARY ESSENTIAL TREMOR

Twenty index patients with hereditary essential tremor, and 93 first degree and 38 more distant relatives were studied at the Institute of Neurology, Queen Square, London, and Oldchurch Hospital, Romford, UK. Tremor presented in the arms. It was symmetrical in 75% and first noted in the dominant hand in 25%. In index patients, tremor spread to affect the legs, head, voice, tongue, face, and rarely the jaw. Affected relatives had tremor of the upper limbs, and spread occurred in a minority. The median age of onset for index patients was 15 yrs (range 5 - 52). The age at onset for both index and secondary cases was bimodal, peaking in the second and fifth and sixth decades. Of 14 relatives under 15 years old, 4 were described as tremulous and 2 had definite tremor, beginning as early as 2 years of age. Disability occurred in the majority but none before 15 years of age. Hunger, emotion, fatigue, and heat exacerbated tremor and disability, whereas alcohol was of benefit in 50%. Classical