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NEUROFIBROMATOSIS

CNS TUMORS AND NEUROFIBROMATOSIS

The association of CNS tumors and neurofibromatosis was studied in a population-based-case control group of 338 patients, less than 15 years of age, diagnosed with a primary tumor of the central nervous system in a ten year period in 53 New York State counties and reported from the Cancer Etiology Unit, Division of Epidemiology, New York State Department of Health, Corning Tower, Albany, NY. The study included 676 controls and information on neurofibromatosis and congenital anomalies. The study confirmed the strong association of neurofibromatosis with the risk of CNS tumors. Thirteen cases and no controls had neurofibromatosis. The types of CNS tumor were optic glioma in 9 patients, astrocytoma in 2, brain stem glioma in 1, and ependymoma in 1. Two fathers and three mothers of these cases had neurofibromatosis and five cases had siblings with neurofibromatosis. Seizures occurred in 37 (12% of 301) cases and in 18 (2.7% of 658) controls. The relative risk of seizures was 4.49 among patients with CNS tumors. There was no difference between cases and controls in the occurrence of congenital anomalies. (Baptiste M et al. Neurofibromatosis and other disorders among children with CNS tumors and their families. Neurology April 1989; 39:487-492).

COMMENT. Neurofibromatosis carries an increased risk of CNS tumors including optic gliomas, other astrocytomas, acoustic neurinomas and meningiomas, also neuroblastoma, leukemia and other cancers. The coincidence of optic glioma and neurofibromatosis is especially frequent in children under 10 years of age and in this study the majority of the CNS tumor cases with neurofibromatosis were optic gliomas. Neurofibromatosis is due to an autosomal dominant genetic mutation with 100% penetrance. An important role for genetics in the etiology of some CNS tumors in children is supported by the strong association of neurofibromatosis with the occurrence of CNS tumors and the excess of neurofibromatosis in parents of CNS tumor cases. Children with

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neurofibromatosis should be examined at regular intervals for possible optic glioma and those with optic glioma should be evaluated for manifestations of neurofibromatosis.

LEARNING PROBLEMS AND NEUROFIBROMATOSIS

A group of 32 children with von Recklinghausen's neurofibromatosis (VRNF) and school learning problems was compared to a matched sample of learning-disordered (LD) students without known genetic or medical disorders at the Division of Psychology, Department of Pediatrics, University of Iowa, Iowa City. VRNF children were found to differ from LD children on the frequency of WISC-R verbal-performance IQ discrepancies, perceptual organization, and on specific measures of visual-perceptual functioning. VRNF children were more likely to display nonverbal learning problems that may affect written language and organizational skills. Only 4% of the VRNF group showed pure language or memory deficits compared to 60% of LD children. Conversely, VRNF children were more likely to present with isolated visual perceptual deficits than LD children (56% versus 6%). The VRNF population could be divided into distinct subgroups: 1) those with no cognitive disabilities; 2) those with mixed language and nonverbal dysfunctions (overlapping with LD children in the general population), and 3) those with nonverbal disabilities (visual perceptual deficits or more subtle organizational difficulties). The majority of VRNF children displayed impulsivity and social imperception, two behavioral features of visual perceptual disability. Eight children (25% of the group) with VRNF showed CNS complications and these children with CNS involvement had slightly lower IQ and cognitive measure scores than the uncomplicated VRNF children. The lower levels of performance may have been due to tumor variables or treatment variables (surgical intervention, cranial irradiation, or anticonvulsant medication). (Eliason MJ. Neuropsychological Patterns: Neurofibromatosis compared to developmental learning disorders. Neurofibromatosis Jan-Feb 1988; 1:17-25).

COMMENT: The learning problems of children with VRNF have some similarities but some differences compared to children in the general population with learning disabilities. The similarities include a preponderance of affected males in both groups, average full scale IQ, and the presence of specific cognitive deficits. The differences include the high frequency of nonverbal dysfunctioning in the VRNF group, the less severe reading deficits in VRNF, and a greater reported frequency of behavior problems. The behavioral symptoms associated with visual perceptual disorder, speech impediments, and the cosmetic disfigurement among children with VRNF may all lead to a general impression of lower intelligence. Earlier studies of individuals with VRNF reported a high frequency of mental retardation and psychiatric disturbances, suggesting that 25-40% of individuals with VRNF were mentally deficient. More recent reports have included less severely affected individuals and the frequency of mental subnormality was closer to 8%. The rate of mental retardation was related to age: It was highest (17%) in the infant toddler group (based on the Bayley Scales), 11% in the preschool group (McCarthy Scales), 7% in the childhood adolescence group (WISC-R), and only 3% in the adult group (WAIS-R).