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NEUROFIBROMATOSIS

OPTIC PATHWAY TUMORS AND NF-1

The natural history of 33 optic pathway tumors (OPT) diagnosed in 227 children with neurofibromatosis type 1 (NF-1) seen in a specialty clinic was evaluated in a prospective, longitudinal study at Children's Memorial Hospital and Northwestern University Medical School, Chicago. OPTs were found in 19% of 176 who had CT or MRI at a median age of 4.2 years. No OPT developed on later follow-up in children who had not received an MRI. In those without ophthalmological complaints at diagnosis, the incidence of OPT was 15%. The median age of children with ophthalmologic complaints such as proptosis or glaucoma (1.9 years) was significantly lower than that of children without such complaints (5.3 years). Only 8 tumors were discovered because of visual complaints or precocious puberty; 25 (76%) children were asymptomatic at time of diagnosis, and 21 (64%) had normal eye findings. Six with chiasmal tumors had decreased visual acuity without proptosis. At follow-up of 0.2 - 8 years, only 3 (9%) showed progressive tumor growth on MRI or deteriorating vision after diagnosis. Symptomatic OPT were diagnosed before the age of 6 years. (Listernick R, Charrow J, Greenwald M, Mets M. Natural history of optic pathway tumors in children with neurofibromatosis type 1: A longitudinal study. J Pediatr July 1994;125:63-66). (Reprints: Robert Listernick MD, Div Gen Acad & Emergency Peds, Children's Memorial Hospital, 2300 Children's Plaza, Chicago, IL 60614).

> COMMENT. The child with NF-1 is at greatest risk for symptomatic optic pathway tumor during the first 6 years of life. Tumor growth after 6 years is unusual. Progressive abnormalities and precocious puberty occurred only with chiasmatic tumors. Tumors confined to the optic nerve discovered by screening with MRI were not accompanied by decreased visual acuity or evidence of progression. The authors advocate serial ophthalmologic examinations in young children with neurofibromatosis type 1, but find that serial MRI is of limited value in asymptomatic patients. The frequency and indications for neurological and neurosurgical consultations in these patients would be of interest.

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