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OPTIC PATHWAY GLIOMAS IN NEUROFIBROMATOSIS TYPE 1: A MORE FAVOURABLE PROGNOSIS

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Neurofibromatosis type1 (NF1) is an autosomal common desorder associated with more complications of which Optic Pathway Gliomas (OPG)are the predominant intracranial neoplasms .
Our aim was to evaluate the prevalence and the outcome of optic pathways gliomas in NF1 children.
The study included 159 pts with NF1 (94 males and 65 females, aged 1-20 years) followed up for a mean of 6 years (2-15 years). All children were evaluated with a detailed physical, neurological and ophthalmological examination. Children with ophthalmological abnormalities and/or precocious puberty underwent a MRI of the head and orbits with contrast enhancement.
Eight children showed symptoms such as endocranic hypertension, seizures, headache i.e., four patients showed only ophthalmological anomalies; two patients had no symptoms or signs.
All children had evidence of OPG on MRI. Three had a prechiasmal tumour, two had a chiasmal tumour, one had prechiasmal/chiasmal chiasmal tumour, two had a pre-chiasmal/ chiasmal and post-chiasmal tumour, two had a chiasmal and post-chiasmal tumour, four had a massive involvement of the optic system. In 4 pts a partial and/or subtotal regression was detected, one child showed evidence of progressive disease.
Because OPG arise in children young than 7 years of age, all NF1 children should undergo yearly to ophalmologic evaluations and annual assessments of growth to monitor for signs of precocious puberty. In the presented series the benigne course of gliomas in NF1 children is confirmed.
Our data confirm the indolent nature of OPG in children with NF1: only 28.6% patients showed progression of disease.