Albinism: diagnosis by visual evoked potentials

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BACKGROUND: In albinism the majority of the nerve fibers of the optic nerve originating from an eye are innervating the contralateral hemisphere. As a result of the predominantly monocular innervation of the left and right hemispheres, the unilateral activation of the visual cortex (lateralization) can be detected with visual evoked potentials (VEP). PATIENTS AND METHODS: The VEPs were elicited with pattern onset stimulation. Five channels were recorded (2 channels over the left and right occipital lobe and 1 in the midline). 22 Patients with pendular nystagmus and 15 normal volunteers were examined. 5 of the normal volunteers were also examined with simulated pendular nystagmus (5-10 degrees, 1 Hz). The nystagmus was simulated by a simple galvanometer scanner based optical system. RESULTS: Unilateral activation of the hemispheres (lateralisation) was never detectable by normal volunteers. The lateralization of the VEP was detectable in all patients with oculocutaneous albinism with pattern onset stimulation. CONCLUSION: The pathological crossing of the nerve fibres in the optic chiasm is always detectable in oculocutaneous and ocular albinism with multichannel pattern onset VEP. The lateralization of the VEP is a more sensitive indicator of ocular albinism than other symptoms like macular hypoplasia, hypopigmentation, iris transillumination, nystagmus, reduced visual acuity et cetera.

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