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### **12.01 Visual evoked potentials in patients with inherited optic neuropathy**

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**Purpose:** In patients presenting with visual loss, visual evoked potential (VEP) responses enable us to evaluate optic nerve function. The purpose of this study was to analyze VEP responses in a series of patients with inherited optic neuropathy referred to a neuro-ophthalmologic clinic.

**Methods:** Patients referred between January 1st 2016 and December 2017 with inherited optic neuropathy were retrospectively included. In all patients undergoing VEP, the electrophysiological results were compared with clinical examination and ocular coherence tomography findings. Flash and pattern VEP recording with different check sizes (60', 30', 15', 7') was performed.

**Results:** Sixty-eight patients met the inclusion criteria and 27 had undergone flash and pattern VEP on presentation. On clinical examination, the optic disc was noted to be either as pale or unremarkable by the referring ophthalmologist. Two different situations were encountered: (i) in 7 patients (13 eyes) with Leber's Hereditary Optic Neuropathy (LHON) who presented with an acute visual loss in at least one eye, no pattern VEP responses were obtained and the temporal RNFL thickness was normal or increased; (ii) in 20 patients, the visual loss was slowly progressive with two subgroups regarding the VEP results: in 21 eyes, an undetectable pattern VEP P100 response was associated with a reduced mean temporal RNFL of 37.1  $\mu\text{m}$  (range 21–43); in 19 eyes, a P100 response could be obtained (at least with a 60' check) and the mean temporal RNFL thickness was also reduced: 40.7  $\mu\text{m}$  (range 22–60). The difference in temporal RNFL thickness was not statistically significant ( $p = 0.12$ ). The genetic testing in these patients enabled us to diagnose either autosomal dominant optic neuropathy or slowly progressive LHON.

**Conclusions:** Visual evoked potential is helpful in objectively demonstrating vision loss in patients with a normal or a subnormal appearance of the optic disc and normal RNFL thickness as encountered in acute LHON. In progressive visual loss such as autosomal dominant optic neuropathy, the VEP responses enabled us to evaluate the functional status which appears to be independent of the temporal RNFL thickness.