Mitochondrial dysfunction affecting visual pathways

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Résumé en anglais	Mitochondrial dysfunction leads to cellular energetic impairment, which may affect the visual pathways, from the retina to retrochiasmal structures. The most common mitochondrial optic neuropathies include Leber's hereditary optic neuropathy and autosomal dominant optic atrophy, but the optic nerve can be affected in other syndromic conditions, such as Wolfram syndrome and Friedreich's ataxia. These disorders may result from mutations in either the mitochondrial DNA or in the nuclear genes encoding mitochondrial proteins. Despite the inconstant genotype-phenotype correlations, a clinical classification of mitochondrial disorders may be made on the basis of distinct neuro-ophthalmic presentations such as optic neuropathy, pigmentary retinopathy and retrochiasmal visual loss. Although no curative treatments are available at present, recent advances throw new light on the pathophysiology of mitochondrial disorders. Current research raises hopes for novel treatment of hereditary optic neuropathies, particularly through the use of new drugs and mitochondrial gene therapy.
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Liens

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[2] http://okina.univ-angers.fr/patrizia.bonneau/publications
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