Vision deficits precede structural losses in a mouse model of mitochondrial dysfunction and progressive retinal degeneration

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Current animal models of retinal disease often involve the rapid development of a retinal disease phenotype; however, this is at odds with age-related diseases that take many years to manifest clinical symptoms. The present study was performed to examine an Apoptosis-inducing factor (Aif) -deficient model, the harlequin carrier mouse (X(hq) X) , and determine how mitochondrial dysfunction and subsequent accelerated aging affect the function and structure of the mouse retina. Vision and eye structure for cohorts of 6 X(hq) X and 6 wild type mice at 3, 11, and 15 months of age were studied using in vivo electroretinography (ERG) , and optical coherence tomography (OCT) .

Retinal superoxide levels were determined in situ using dihydroethidium (DHE) histochemistry. Retinal cell counts were quantified post mortem using hematoxylin and eosin (H&E) staining. ERG analysis of X(hq) X retinal function indicated a reduction in b-wave amplitude significant at 3 months of age (p
http://www.ncbi.nlm.nih.gov/pubmed/21983042