

# OPHTHALMIC FINDINGS IN CEREBELLAR ATAXIA, NEUROPATHY AND VESTIBULAR AREFLEXIA SYNDROME (CANVAS)

**Purpose:** Cerebellar ataxia, neuropathy and vestibular areflexia syndrome (CANVAS) was recognised as a distinct clinical syndrome in 2011. Afferent visual system abnormalities are an important feature of other ataxias but have not been studied in CANVAS. The aim of this study was to characterise the ophthalmic features of the condition.

**Method:** Neuro-ophthalmic and neurological examination, including optical coherence tomography (OCT) scans of the macula and peripapillary retinal nerve fibre layer (RNFL), was undertaken for 16 CANVAS participants and 15 age-matched healthy controls.

**Results:** Mean age at symptom onset was  $53.2 \pm 8.4$  years. Nystagmus was observed in all patients. Visual acuity was reduced in the CANVAS group (logMAR  $0.17 \pm 0.15$  vs.  $0.00 \pm 0.04$  in controls,  $p = 0.0002$ ). There was a significant reduction in temporal RNFL thickness in the CANVAS patient group compared with controls ( $58.8 \pm 6.1 \mu\text{m}$  vs.  $71.7 \pm 11.6 \mu\text{m}$ ,  $p = 0.005$ ). There were no significant differences between the two groups in other OCT measures. There were no significant relationships between ophthalmic findings and measures of disease severity.

**Conclusion:** This is the first report to detail afferent visual system findings in CANVAS. Temporal RNFL thinning is a feature of several mitochondrial disorders and this requires further investigation in CANVAS. The primary limitation of this study was the limited size of the CANVAS cohort, due to the relative rarity of the disease. Larger, longitudinal studies are required to further clarify the clinical relevance of this finding.