

Cerebellar Ataxia with Auditory Neuropathy And Vestibular Areflexia. A New Variant of CANVAS Syndrome?

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Objectives: None of the previous studies highlighted the occurrence of hearing loss with intact sensory and motor functions in CANVAS, accordingly, we present here a case report of a unique type of hearing loss in addition to absence of sensory or motor neuropathy.

Background: Patients with a combination of bilateral normal peripheral hearing with selective bilateral vestibulopathy and cerebellar ataxia were first described in 1990.

Methods: Bedside vestibular examination and monothermal ice caloric test.
Brain MRI
Pure tone audiometry
Otoacoustic emissions.

Results: ANSD

Conclusion: Assessment of vestibular function in addition to cochlear function using ABR and OAEs should be performed to look for other variants of CANVAS cases