

NEUROLOGY

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Neurology 2008;70;1217-1218

DOI: 10.1212/01.wnl.0000307752.25133.8f

This information is current as of September 10, 2008

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PULSE-SYNCHRONOUS TORSIONAL PENDULAR NYSTAGMUS IN UNILATERAL SUPERIOR CANAL DEHISCENCE



A physically active 62-year-old man experienced the gradual onset of intermittent positional dizziness, occurring primarily on inclining his head upwards or downwards. Dizziness also appeared less commonly with abrupt turns of the head or while bearing down during a bowel movement. He denied tinnitus. He had no significant family history, he had never sustained head or neck trauma, and he was on no medications. Treatment with meclizine had not alleviated his symptoms.

On physical examination with video Frenzel goggles (Micromedical Technology, Chatham, IL), there was a primary position torsional nystagmus, of approximately 1 Hz. There was also a low-amplitude downbeating nystagmus, greater on lateral gaze, and with no positional modulation. By watching the nystagmus while palpating the pulse, it was appreciated that the torsional nystagmus was pulse-synchronous, with the clockwise component corresponding to the pulse upswing. The torsional nystagmus was present when the patient was sitting or standing, but was suppressible in those positions with the Valsalva maneuver, and was absent when supine (see video on the *Neurology*[®] Web site at www.neurology.org).

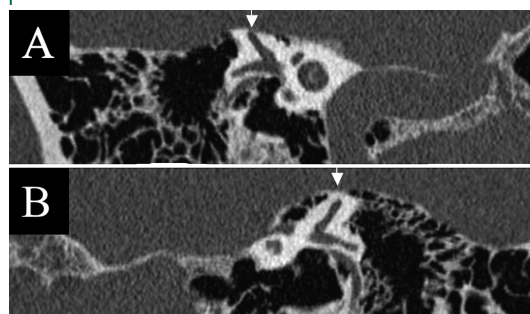
Laboratory investigation revealed vestibular evoked myogenic potentials to be fourfold enlarged on the right, and the responses could be obtained at a lower threshold on the right. Audiometry demonstrated reduced hearing, with bone conduction being better than air, on the right. Electronystagmography revealed that the caloric responses on the right were 54% weaker than the left. Electrocochleography was normal. MRI of the brain was unremarkable. High resolution CT imaging revealed superior canal dehiscence on the right side (figure).

Superior canal dehiscence (SCD) is a recently described disturbance of the inner ear in which there is a defect in the bony roof of the superior semicircular canal.¹ The defect is covered by dura,^{2,3} so there is no direct communication of fluid between the CSF and the perilymphatic space. Although the etiology is unknown, a large case series of temporal bones³ revealed that the bone overlying the superior canal is thin (<0.1 mm) in 1.3% of individuals, which has led to speculation that in such individuals minor head trauma or an abrupt change in intracranial pressure (as in sneezing) may induce frank bony dehiscence. SCD typically presents with sound-induced dizzi-

ness (Tullio's phenomenon), and provocative maneuvers usually produce a mixed torsional and vertical nystagmus consistent with stimulation of the superior canal.⁴ Pulse-synchronous torsional pendular nystagmus in association with unilateral SCD has been reported previously.^{2,5} Our report adds to these data and also observes that in this case the nystagmus was suppressible by the Valsalva maneuver or by lying supine.

We postulate the following as a mechanism for this finding. Systemic arterial pulse pressure is transmitted intracranially, causing pulse-synchronous fluctuation in intracranial pressure. In SCD, intracranial pressure fluctuations may be transmitted from the intracranial compartment by CSF pushing against the dura² overlying the defect in the bony roof of the superior canal, which in turn induces movement of the perilymphatic fluid and endolymphatic fluid⁶ in the superior canal, causing nystagmus and dizziness. The two maneuvers that we observed to eliminate the pendular nystagmus both increase intracranial pressure; the Valsalva maneuver or lying supine. To explain this patient's findings, we hypothesize that lower baseline intracranial pressure permits the dura overlying the bony defect to transmit pressure fluctuations from the intracranial compartment to the perilymphatic space of the superior canal, resulting in ocular torsion. In contrast, higher intracranial pressure associated with supine position or with Valsalva maneuver may make the dural compartment less compliant, and less able to transmit pressure fluctuations. Although the presence of a component of low-amplitude downbeating nystagmus raises the possibility of a central lesion, MRI of the brain demonstrated no abnormalities.

Figure High-resolution CT images of the temporal bones



Collimated axial images (1.5 mm) were obtained, from which were reconstructed images parallel and perpendicular to the superior canal, and in the coronal plane. (A) Right temporal bone, demonstrating dehiscence of the superior canal (arrow). The diameter of the defect measures approximately 1.3 mm. (B) Left temporal bone, demonstrating the superior canal without dehiscence (arrow).

Supplemental data at
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Torsional pendular nystagmus is most commonly reported to be caused by brainstem lesions. These mechanisms are due to central oscillators that produce eye movement without an external trigger. Our case demonstrates that SCD is a mechanical, peripheral vestibular etiology that should be considered in the differential diagnosis of torsional pendular nystagmus. Furthermore, it can be distinguished from central etiologies of torsional pendular nystagmus by observing pulse synchronization and modulation by maneuvers that change intracranial pressure. Other potential etiologies of peripheral torsional pendular nystagmus include venous malformations,⁷ fistulae, and posterior canal dehiscence.

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Disclosure: The authors report no conflicts of interest.

Received September 22, 2006. Accepted in final form August 8, 2007.

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INTERMITTENT PREDNISOLONE AND AUTOANTIBODIES TO GAD65 IN JUVENILE NEURONAL CEROID LIPOFUSCINOSIS

Juvenile neuronal ceroid lipofuscinosis (JNCL) is a recessively inherited, progressive neurologic disease caused by mutations in the *CLN3* gene, coding for a transmembrane protein with unknown function.¹ The symptoms include visual failure, noticed around the age of 4 to 7 years, mental decline, epilepsy, parkinsonism, psychiatric symptoms, and sleeping difficulties. Thus far, no curative treatment is available and the disease leads to an early death at the age of 15 to 30 years.

Autoantibodies to glutamic acid decarboxylase 65 (GAD65) were detected in the sera of patients with JNCL and in *CLN3* knock-out mice.² As a biologic sign of these antibodies, decreased GAD65 activity and increased levels of glutamate were observed in the mice brains. Together these findings suggest that an immune-mediated reaction to GAD65 may be involved in the pathogenesis of JNCL.

Our aim was to test the effect of immunosuppressive therapy on the GAD65-antibodies, symptoms, and disease progression in patients with JNCL.

Methods. Four girls and four boys with JNCL (table) were included in this 1-year study, ap-

proved by the Ethics Committee of the Hospital for Children and Adolescents, University of Helsinki.

The patients visited the outpatient clinic every 6 months and their motor function was scored using the motor part of the Unified PD Rating Scale (UPDRS).³ At the beginning and end of the study, the patients underwent a neuropsychological examination including the assessment of verbal IQ.⁴ Relative height (SD score) and weight for height (%) were assessed from Finnish growth charts.⁵ The scores for UPDRS, relative height, and weight for height were compared at the beginning of the study and 1 year later using the Wilcoxon Signed Rank Test. The change in IQ was studied using the Reliable Change Index.

Levels of GAD65-antibodies in the patients' sera were studied using a recombinant human GAD65 protein as a probe in Western blot and dot-blot analysis. The patient sera (collected at 0, 3, 6, and 12 months) served as primary antisera. GAD65-antibodies were visualized with peroxidase-conjugated anti-human IgG and chemiluminescent substrates. Serum from a patient with stiff-man syndrome served as a positive and serum from healthy subjects as negative controls.

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