

TITLE: Superior Canal Dehiscence
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Introduction:

Superior Canal Dehiscence has recently been described as an etiologic agent in vertigo.

History:

Superior Canal Dehiscence as a cause of sound and/or pressure-induced vertigo was first postulated in 1998, in a case series described by Minor *et al*¹. This study described eight patients with vestibular symptoms (oscillopsia, vertigo), as well as chronic disequilibrium. All patients were found to have evoked eye movements with vertical and torsional components, with the direction corresponding to inhibition or excitation of the affected semicircular canal. Axial and coronal computed tomography (CT) of the temporal bone and labyrinth revealed dehiscence of the bone overlying the superior circular canal. A study by Carey *et al*² studied the incidence of superior canal dehiscence in a temporal bone archive. Minimum thickness of the bone was measured at the floor of the middle cranial fossa, and at the superior cranial fossa. Complete dehiscence was identified in 0.5% of specimens, and thinning (<0.1mm) of the bone in an additional 1.4%. Thinning of the bone could also be detected on ultra-high-resolution CT scan and identified as dehiscent. Individuals with dehiscence or bone thinning over the superior canal are thought to be prone to development of superior canal dehiscence syndrome with its associated clinical symptoms if trauma disrupts the fragile bone¹. Minor *et al* reported that up to 23% of patients describe an inciting event – loud noise, heavy lifting, trauma to head – precedes the first symptomatic episode of superior canal dehiscence.

Embryologic Development:

At 15 weeks of gestation, ossification centers of the ear grow and fuse such that by 21 weeks, the bony labyrinth encases the membranous labyrinth. Each ossification center has a trilaminar structure: The outer periosteal layer continues to grow postnatally. The middle layer forms by invasion of osteoblast into cartilage, and eventually forms the petrous portion of the temporal bone. The innermost periosteal layer contacts the perilymphatic space, and overlies the superior canal where it enters the middle cranial fossa. Postnatal failure to develop outer/middle layer bone to cover the periosteal layer over the semicircular canal has been suggested as the cause of dehiscence. The other hypothesis, that erosion of the bone occurs gradually over the individual's lifespan, is not supported by histologic evidence as ossification patterns appear stable over time rather than altered² by an acute event.

Anatomy and Physiology:

The semicircular canals play an essential role in balance and perception of movement. They consist of three membranous interconnected tubes shaped as a torus, positioned orthogonally to each other. The membranous portion of the canal is attached to temporal bone. The ampulla is located at one end of the torus, with a gelatinous cupula dividing one end of the ampulla from the other. Each canal is filled with endolymph and contains cilia embedded in the cupula. Maximum sensitivity of the canal is rotation in the plane of the torus³, with the horizontal canal detecting horizontal head movements, while the posterior and superior canals detect motion in the vertical direction.

The fluidity of endolymph allows it to move within the semicircular canal; however, its inertial property causes it to initially oppose the turning motion. The elastic component of the cupula allows it to detect slight changes in pressure and deflect accordingly. As the cupula deflects, stereocilia move toward or away from the kinocilium, stimulating or inhibiting the vestibular nerve, respectively. Kinocilia are positioned in a parallel fashion to the long axis of each semicircular canal; in the posterior and superior semicircular canal, ampullofugal flow of endolymph is excitatory as the hair cells are organized so that kinocillia are located further away from the vestibule. In the horizontal canal, the excitatory direction is reversed as the kinocillia are closer to the vestibule. Thus, ampullopetal flow stimulates vestibular nerve firing in the horizontal canal.

Pathophysiology:

Superior canal dehiscence syndrome results when the temporal bone overlying the superior circular canal is absent. Under normal conditions, sound pressure enters the inner ear via the stapes footplate and oval window, rotates through the cochlea and exits via the round window⁴. In superior canal dehiscence, the lack of bone overlying the area results in greater compliance⁵, creating a third window space and thus allowing abnormal movement of endolymph within the canal in the presence of certain stimuli. These stimuli increase the pressure gradient within the membranous labyrinth: stimuli that result in inward pressure at the round or oval windows (such as Valsalva against closed nostrils, or tragal compression) cause outward bulging at the dehiscence site⁶, with resulting ampullofugal deflection of superior canal cupula. Inward bulging

at the bony dehiscence results in ampullopetal deflection of the cupula; this can be caused by Valsalva against a closed glottis or jugular venous compression.

Evoked eye movements that are seen in patients with superior canal dehiscence syndrome are related to Ewald's first law: stimulation of the superior semicircular canal produces eye rotation in the vertical-torsional plane. Torsional movements are counterclockwise if canal dehiscence is seen on the right, clockwise if it is present on the left⁵.

Clinical Findings and Diagnosis:

Superior canal dehiscence is a primarily clinical diagnosis. Suspicious historical findings include Tullio's phenomenon, which is dizziness/vertigo induced by sounds, and oscillopsia, a visual disturbance where objects in the field of vision seem to oscillate. Autophony, where the patient reports hearing their own voice louder in the affected ear, is another common finding in this disorder. When hearing loss is present, it is a conductive hearing loss secondary to dissipation of acoustic energy transmitted through air conduction – bone conduction is increased in patients suffering from superior canal dehiscence. This increased bone conduction is apparent on physical exam, where Rinne test will be negative in the affected ear. Weber test will lateralize toward the affected ear, which is also consistent with the finding of conductive hearing loss. Vestibular testing shows some variability among subjects.

However, most have a negative Dix-Hallpike test (in the absence of concurrent benign paroxysmal positional vertigo). Videonystagmography (VNG) testing, Frenzel goggles, tragal compression or pneumatic otoscopy can all cause eye movements in the plane of the superior canal as well as concurrent dizziness. An audiometer fitted to patient during evoked eye movements can also show nystagmus to low-frequency, high-intensity sounds. Though vestibular testing can aid in the confirmation of a vertiginous disorder, the gold standard of diagnosis of superior canal dehiscence is a high resolution 0.5 mm collimation or less CT of the temporal bones with images that are reformatted to include cuts in the plane of (Pöschl), as well as perpendicular (Stenver) to, the superior canals.

Medical and Surgical Interventions:

Severity of a patient's symptoms and impact of these symptoms in the patient's daily life determine the degree of intervention. Patients who are found to have dehiscence of the superior canal as an incidental finding on a CT scan can be followed until such a time as symptoms develop. Patients with mild symptoms can be managed conservatively with avoidance of causative stimuli. Myringotomy tubes can be placed in patients with primarily pressure-related symptoms. The decision to insert surgical plugging of the canal to alleviate symptoms should be made based on symptoms and patients' comorbid conditions as well as ability to undergo surgical procedure.

Current surgical options include canal resurfacing and canal occlusion. The site of dehiscence is located on the medial aspect of the superior semicircular canal, which contacts the superior petrosal sinus or, less frequently, the middle ear fossa⁴. Procedure can enter through transmastoid approach, or via middle fossa craniotomy. Dural elevation over arcuate eminence to avoid damage to membranous labyrinth is performed, and dehiscence is either closed with fascia

and bone dust or sculpted cortical bone, or canal is occluded completely with bone paté. Superior canal resurfacing has the advantage that it preserves the physiologic function of the superior semicircular canal, but may result in recurrence should the fascia or bone graft move out of place. Canal plugging has a lower rate of recurrence, but results in more trauma to the membranous labyrinth.

Current Research:

The surgical approach to superior canal dehiscence continues to be in development, with no long-term follow-up studies available to evaluate benefit to risk ratio of canal plugging vs. canal resurfacing at this time. Canal plugging is currently the most used procedure due to its use by the surgeons who first described the syndrome. Another area of research delves into whether the size of the dehiscence will impact symptom severity; in a study by Pfammater *et al*⁷, results showed that patients with large dehiscences have significantly increased vestibulocochlear symptoms, lower VEMP thresholds, and increased rate of objective vestibular findings as compared to patients with smaller dehiscences.

In summary,

Superior Canal Dehiscence is a relatively new diagnosis in the field of Otolaryngology. It is characterized by pressure and/or sound-induced vertigo, evoked eye movements in the plane of the affected semicircular canal, as well as increased bone conduction and conductive hearing loss. Medical and surgical management is continually evolving as more accurate understanding of the syndrome emerges.

Discussant's Remarks: Tomoko Makishima, M.D

This was a well designed, concise review on a relatively new disease entity, "superior canal dehiscence". I believe that in most cases, the presenting symptoms and signs are very subtle in the beginning. Therefore, the key to diagnosis is to suspect "superior canal dehiscence". Now that is more widely known, more and more cases are being reported, which will in turn enrich our knowledge base on this condition. The treatment options and the accompanied prognosis is still mostly uncertain. This presentation covered most of what is known today. I am looking forward to revisit this topic again in a few years.

References

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