

## Superior semicircular canal dehiscence syndrome

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### Abstract:

This article discusses superior semicircular canal dehiscence syndrome. This syndrome has been identified in 1998 by Minor et al. This condition is caused due to dehiscence of bone overlying the superior semicircular canal. This causes vertigo, oscillopsia, dysequilibrium due to exposure to sound. Ultra high resolution CT scans help in identification of this condition. This dehiscence has been confirmed by surgical exploration of middle cranial fossa.

### Introduction:

Superior semicircular dehiscence syndrome is a reasonably recently identified condition. It was Minor et al<sup>1</sup> during 1998 who documented first this condition. This condition causes sound / pressure induced vertigo, autophony, conductive hearing loss and hyperacusis in some patients. Sound induced vertigo also known as Tullio phenomenon<sup>2</sup> classically seen in this condition can also be caused by perilymphatic fistula, congenital syphilis (due to excessive laxity of annular ligament).

### Clinical features of Superior semicircular canal dehiscence syndrome:

1. Sound / pressure induced vertigo (Tullio phenomenon)<sup>3</sup>
2. Nystagmus is vertical and rotatory in nature. This is in line with the involved superior semicircular canal. This may vary in patients with large dehiscence over superior canal due to hyperfunction.
3. Vertigo during coughing, sneezing and valsalva manoeuvre. This is due to increased middle ear pressure.

### Pathophysiology:

Under normal circumstances the round and oval windows are the only two openings seen in the hydraulic system of inner ear. Inner ear corresponds to a hydraulically closed system. There is no substantial movement of inner ear fluid in the semicircular canal even when stapes vibrates in response to sound. When a third window is present in this system in the form of labyrinthine fistula then membranous labyrinth over the fistulous area bulges out causing movement of endolymph.

This affects the hydraulic purity of the system causing the brain to interpret this movement as movement of the body. This causes a conflict with input received from visual and proprioceptive receptors leading on to a sense of reeling by the patient.

Presence of third window in the labyrinth is the cause for positive Tullio phenomenon. According to Minor lack of bony covering over superior semicircular canal could make it to function like a third window in addition to round and oval windows.

Incidence:

The exact incidence of superior semicircular canal dehiscence syndrome is currently not known. According to Carey et al who studied nearly 1000 cadaver temporal bones estimated complete dehiscence of bony covering over superior semicircular canal to be about 0.5%.<sup>4</sup> This study also estimated thinner covering of bone over superior semicircular canal in 1.5% of bones studied. In infant temporal bones the bony covering over superior semicircular canal is very thin and undergoes progressive thickening till the child reaches the age of 3. This same phenomenon had been observed in dogs too.<sup>5</sup>

Other associated abnormalities noticed along with superior semicircular canal dehiscence:

1. Tegmen dehiscence. This is more common in patients with superior semicircular canal dehiscence than normal population<sup>6</sup>.
2. Absence of bone covering over geniculate ganglion. This is higher than that of normal population.<sup>7</sup>

Clinical presentation:

These patients present with episodes of vertigo and oscillopsia which are induced by three possible factors:<sup>8</sup>

1. Loud noises (Tullio phenomenon)
2. Changes in pressure in the external auditory canal transmitted to the middle ear (Hennebert's sign)
3. Valsalva manoeuvre

Conductive hearing loss:

Conductive hearing loss in isolation or along with vestibular symptoms are also seen in these patients. When present in isolation it is commonly mistook for otosclerosis.<sup>9</sup> It should also be

borne in mind that in patients with superior semicircular canal dehiscence syndrome stapedial reflex is normal whereas in otosclerosis it is lost. A study of stapedial reflex will help in differentiating these patients from those suffering from otosclerosis.<sup>10</sup>

House et al in 1980<sup>13</sup> used the term inner ear conductive deafness to describe patients who had persistent conductive hearing loss despite undergoing successful stapedectomy surgery.

#### Diagnosis:

Is made from history and imaging. Imaging is currently widely relied on because it is the only currently available non-invasive investigation available for this condition. High resolution CT imaging coronal cuts 0.5 mm<sup>11</sup> is preferred to eliminate false results due to partial averaging errors. Electronystagmography can be used to record the type of nystagmus and its direction.

#### Vestibular evoked myogenic potentials:

This test has been proved to be useful in the diagnosis of superior semicircular canal dehiscence syndrome<sup>12</sup>. This test is based on recording the short latency relaxation potentials over the skin lining the ipsilateral sternomastoid muscle in response to click stimuli. The threshold to evoke these responses is lowered in these ears when compared to normal individuals.



CT scan temporal bone showing dehiscent superior semicircular canal

Management:

If symptoms were mild then simple repositioning maneuvers could help in alleviating positional vertigo. If symptoms are debilitating then surgery is the option.

Surgery can be performed via middle cranial fossa approach and the dehiscence is sealed with fascia and fast setting bone cement. Surgical repair has a success rate of more than 90%. This highly under diagnosed condition is treatable surgically with excellent cure rates. Even though superior canal can be accessed via conventional mastoidectomy<sup>14</sup>, middle cranial fossa approach is preferred because it allows direct visualization of the dehiscent site.

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