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Seminars in
ROENTGENOLOGY

Imaging of Third Window Lesions of the Temporal Bone

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Physiology of Normal Hearing

The acoustic resistance to passage of sound through a medium is termed impedance. The transduction of vibration from the external environment through the external auditory canal air (low impedance) to the cochlear fluids (high impedance) results from impedance matching function of the middle ear.[1] Three levers help in accomplishing the pressure transformation in the middle ear for transduction of sound vibration.[2] The catenary lever results from the outer convexity of the eardrum and radial orientation of the collagen fibers in the tympanic membrane which lead to a 2-fold amplification of sound pressure onto the umbo. The ossicular lever results from the long axis of the malleus being 1.3 times the length of the long process of the incus. Finally, the hydraulic lever results from a 17-20 fold difference in vibratory surface of the tympanic membrane compared to the stapes footplate. This, in fact is the main impedance matching mechanism in the middle ear and results in a 14 times gain in sound pressure. Normal air conduction of sound (Fig. 1A) occurs via the following mechanism: Sound waves travel from the external environment and are transmitted via the ossicles in the middle ear to the oval window. Sound vibrations are then conducted through the incompressible closed-loop of the scala vestibuli producing equal and outward motion of the round window. The pressure differential between the oval window and round window generates a pressure gradient across the cochlear membrane in the scala media, activating hair cells and creating a perception of sound. Normal bone conduction (Fig. 1C) of sound occurs when sound vibration is transmitted through the otic capsule onto the cochlear membrane in the scala media by direct compression of the bone. This causes squeezing of the perilymph with differential outward motion of the oval window and round window due to unequal impedance of these 2 structures. This inequality of impedance leads to pressure difference across the cochlear partition with resultant motion of the basilar membrane and perception of bone conducted sound.

There are physiological windows in the temporal bone which facilitate hearing.[3] These comprise the oval window, round window, and channels carrying small vessels and nerves. These windows are larger, shorter, have low impedance and facilitate sound transmission. There are additional normal third windows which are smaller and longer channels with high impedance and are functionally closed to sound flow. These include a normal-sized vestibular aqueduct, normal-sized cochlear aqueduct, and foramina for blood vessels and nerves.

The pathologic or third mobile window phenomenon was first described by Merchant et al in 2004.[4] Pathologic third windows like superior semicircular canal dehiscence (SSCD) affect both air conduction and bone conduction in the temporal bone[3] (Fig. 1). The third window created by the dehiscent bone directs air conducted sound energy away from the cochlea elevating air conduction thresholds. There is dampened energy transmission from the oval window to the round window with decrease in pressure gradient across the cochlear membrane and resultant decreased sound perception (Fig. 1B). The dehiscent bone causes decreased motion of the oval window on the scala vestibuli side but the motion of the round window on the scala tympani side is unchanged. This increases the differences in impedance between the scala vestibuli and scala tympani thus improving threshold for bone conducted sound (Fig. 1D). As a result, there is a classic air bone gap on audiogram at low frequencies. At high frequencies, there is a small or no gap since proportionally less acoustic energy is shunted by the third window at these frequencies.

Clinical Features

There is normal or supranormal bone conduction in these lesions as mentioned above. This bony hyperacusis manifest as symptoms like autophony (hearing own voice as loud or distorted), pulsatile tinnitus and audible eye movements, footsteps, chewing, and bowel movements.[5] Conductive hearing loss (CHL) is also seen in these patients due to air bone gap as described above. The hearing loss is also referred to as pseudo CHL since it arises from the inner ear and not from the middle ear which is the usual

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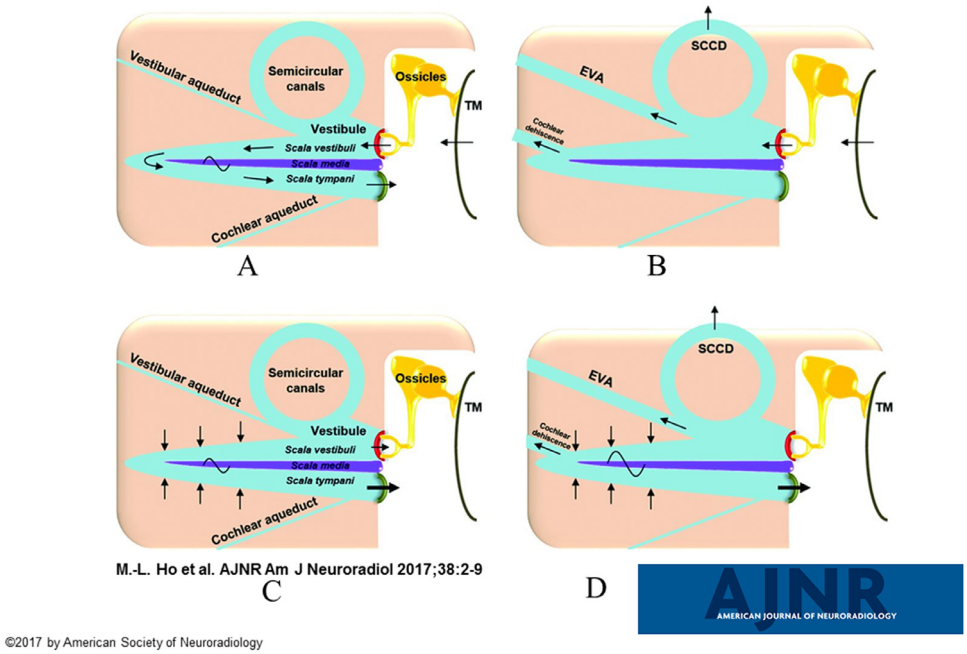

Figure 1 Mechanisms of air- and bone-conducted sound in healthy and third window anatomy. (A), Normal air conduction. (B), Decreased air conduction in third window anatomy. (C), Normal bone conduction. (D), Increased bone conduction in third window anatomy. TM indicates tympanic membrane; yellow, auditory ossicles; beige, otic capsule; red, oval window; green, round window; blue, perilymph; purple, basilar membrane. Adapted with permission from Merchant SN, Rosowski JJ. Conductive hearing loss caused by third window lesions of the inner ear. *Otol Neurotol* 2008;29:282-89. (Color version of figure is available online.)

source of CHL.[6] In patients with SSCD exposed to pressure changes or loud sounds, eye movements are noted in the plane of the ipsilateral superior semicircular canal linking these symptoms to anatomy. The phenomenon of sound induced vertigo and or nystagmus in the plane of the superior semicircular canal is called Tullio's phenomenon. It is postulated that the pressure gradients between the oval window and the dehiscence cause flow of endolymph in the superior canal ampulla contributing to the vertigo and nystagmus. These pressure gradients can also be generated by positive pressure applied to the external auditory canal (Hennebert sign) or valsalva against pinched nostril. Patients also report aural fullness and a sensation of brain fog which may be related to vestibular contributions to cognition.

Disorders Which Result in Mobile or Pathologic Third Window

There are disparate disorders which create a mobile window on the scala vestibuli side of the cochlear partition.[7] These can be classified as focal or diffuse.[3] The most common focal lesion resulting in third window phenomenon is SSCD. Other focal lesions include enlarged vestibular aqueduct, stapes gushers, carotid cochlear dehiscence, and otosclerosis. Paget's disease is a diffuse bone disease which may result in this phenomenon. In general, these lesions demonstrate a normal middle ear on imaging which help differentiate this from CHL resulting from the more common middle ear pathology.

Superior Semicircular Canal Dehiscence

SSCD was first reported by Lloyd Minor et al in 1998.[8] There is a defect in the bone overlying the superior semicircular canal, leading to a communication between the superior semicircular canal and the middle cranial fossa. The true prevalence of this entity is unknown and depends on the methodology used for detection. In a cadaveric study SSCD prevalence was approximately 0.5% of temporal bone specimens (0.7% of individuals). In 1.4% of specimens (1.3% of individuals), the bone was markedly thin (≤0.1 mm) or near dehiscent.[9] In another retrospective study of 500 patients, prevalence on CT of the temporal bone was 2.0% in patients without symptoms of SSCD and 13.6% in patients with symptoms attributable to SSCD.[10]

The etiology of SSCD remains unknown. There is a debate as to whether this is a congenital or acquired lesion.[11] The increased radiologic prevalence of SSCD among older age groups suggests that this is an acquired condition.[12] Osteopenia and chronic otitis media are also associated with SSCD indicating an acquired etiology.[13,14] However, SSCD is also seen in children <2 years (with bone covering of the SSC progressively increasing from 2 to 8 years of age) supporting a congenital cause.[15,16] A study demonstrated increased incidence of bone thinning or dehiscence of the middle cranial fossa floor in SSCD patients compared with normal patients, suggesting that the etiology of SSCD may also affect surrounding structures.[9,17]

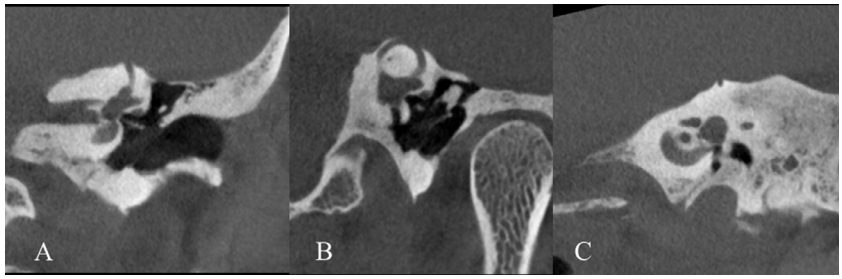

Figure 2 SSCD. Coronal (A), Poschl (B) and Stenver (C) views from a MSCT temporal bone exam reveal a defect in the arcuate eminence at the roof of the SSC.

In about 25% of cases trauma or sudden increase in intracranial pressure can be inciting event.[11] The lesion tends to be bilateral in 25%. Some cases of SSCD found incidentally on imaging without symptoms suggesting a protective role of the dura in plugging the dehiscence and preventing pressure transmission in such individuals.

Imaging diagnosis of SSCD relies on thin multislice CT of the temporal bone. Thin-section 0.5-mm collimation Coronal reformations are sufficient for the evaluation of SSCD (Fig. 2). Additional reformations in the planes of Stenver (perpendicular to the plane of the superior semicircular canal) and Pöschl (parallel to the plane of the superior semicircular canal) improve detectability in our experience although a 2006 study did not advocate their routine use.[18] A radiologic classification of SSCD has recently been proposed.[19] The authors classified the dehiscence by location: the roof/arcuate eminence, lateral upslope, medial downslope, or dehiscence of the superior petrosal sinus into the superior semicircular canal. The most common site of the dehiscence was noted to be at the roof/arcuate eminence followed by the medial downslope. Although some authors have advocated MRI for this diagnosis using volumetric coronal 3D gradient images, MRI cannot conclusively diagnose thin or dehiscent SSC and is not useful in our experience.[20]

Dehiscence of the superior semicircular canal into the superior petrosal sinus is described as a "cookie bite" out of the SSC on CT[21] (Fig. 3).

We have also seen the mobile third window phenomenon in patients who presented with superior canal dehiscence into a bony cyst in the petrous temporal bone (Fig. 4).

Patients with markedly thinned but not frankly dehiscent superior semicircular canals can present with symptoms of SSCD syndrome and may demonstrate low frequency air bone gaps.[22] These patients improve with surgery but may have incomplete symptom relief and possible complications.

The treatment of SSCD include plugging the affected canal by a combination of fascia, bone dust and bone chips.[23-25] Other plugging materials such as bone wax, cartilage, fascia, fibrin glue, and hydroxyapatite cement have also been used. The middle cranial fossa can be resurfaced with fascia and hydroxyapatite cement. Reduction in patient's symptoms is reported regardless of the material used. Recently, reinforcement of the round window has been attempted as a means to dampen the symptoms of SSCD. Although the procedure did benefit some patients, successful outcomes were not predictable and this procedure has not been widely accepted.[26]

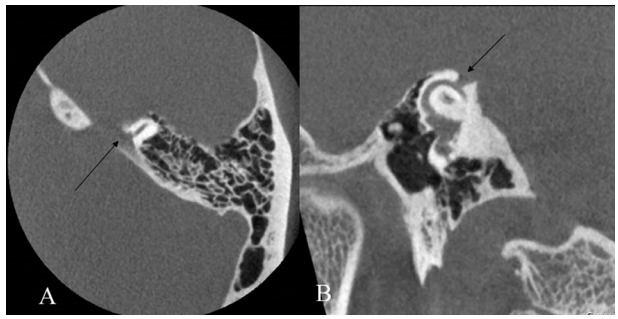

Figure 3 SSCD into superior petrosal sinus. Axial (A) and Poschl (B) view from a MSCT of the temporal bone reveal a characteristic "cookie bite" (arrow) appearance of SSCD into the superior petrosal sinus.

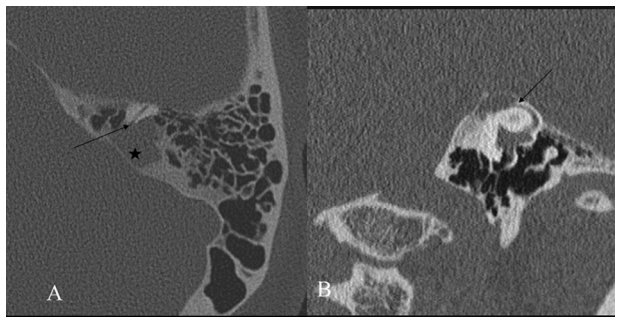

Figure 4 SSCD into bony cyst. Axial (A) and Poschl (B) view from a MSCT of the temporal bone demonstrate the SSC dehiscent (arrow) into a petrous apex cyst (star) in a patient who presented with mobile third window symptoms.

In addition to SSCD, posterior semicircular canal dehiscence can also represent a third-window lesion manifesting as hearing loss with vestibular dysfunction[27,28] (Fig. 5). There are rare cases of idiopathic bony dehiscence at the lateral semicircular canal[29] but most cases of lateral semicircular canal are acquired due to inflammatory disease like cholesteatoma in which the third window phenomenon is usually masked by signs and symptoms of inflammatory disease.

Enlarged Vestibular Aqueduct

Enlarged vestibular aqueduct can also occasionally present with third window phenomenon.[30,3]

The enlarged vestibular aqueduct is postulated to provide pathologically large communication between the bony vestibule and the cranial cavity, resulting in an air-bone gap (Fig. 6).

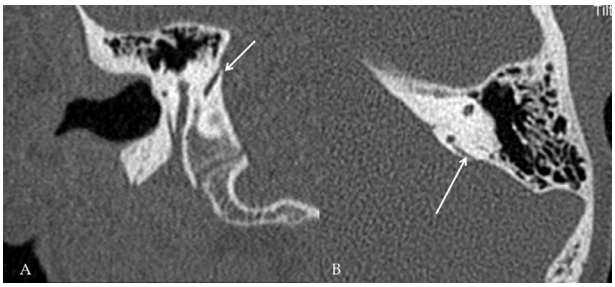

Figure 5 Posterior semicircular canal dehiscence. Poschl (A) and Axial (B) images demonstrate dehiscence of the bone covering the posterior semicircular canal (arrow).

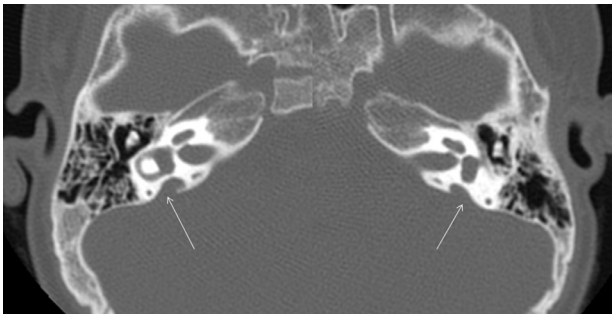

Figure 6 Bilateral EVA. Axial CT images demonstrate bilateral EVA's which can create a pathologic third window. (arrows).

Thin section MSCT of the temporal bone is the modality of choice to diagnose EVA. The Cincinnati criteria define EVA as a vestibular aqueduct with width of 2 mm at the operculum and/or 1 mm at the midpoint as measured on axial images.[31] The 45° oblique plane gives a more reliable depiction of the vestibular aqueduct than the axial plane in CT evaluation of the temporal bone.[32,33]

Miscellaneous Rare Disorders Which May be Associated With Third Window Phenomenon

Jugular bulb-vestibular aqueduct dehiscence has a prevalence ranging from 8 to 11% of patients undergoing temporal bone CT.[34,35] Friedmann et al reported third window symptoms in a small series of case with high riding JB or JBD eroding into the inner ear.[36] However, a follow-up larger study by the same group concluded that most patients with JB associated inner ear dehiscence are asymptomatic.[37] Causative relationship between Jugular bulb-vestibular aqueduct dehiscence and third window phenomenon is still unclear and likely not present.[38,39]

In DFN-3 (X-Linked Deafness with Stapes Gusher) there is an open communication between the inner ear and IAC due to deficiency of intervening bone resulting in a pathologic third window.[3] On CT, there is a characteristic appearance with a dilated IAC, dilated vestibular aqueduct and corkscrew appearance of the cochlea (Fig. 7).

Carotid cochlear dehiscence has been described as a rare cause of third window phenomenon.[40-42] A communication between the cochlea on the scala vestibuli side and the carotid artery is postulated to create a pathologic third window[3] (Fig. 8).

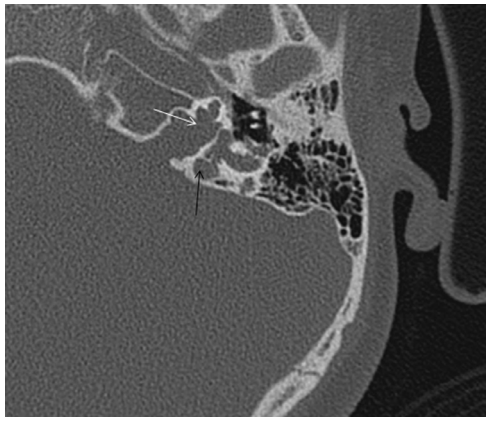

Figure 7 Stapes Gusher. On the axial CT Temporal bone image, there is deficiency of bone between cochlea and IAC (white arrow), the IAC is dilated and there is an EVA (black arrow). This results in a pathologic third window between the cochlea and the IAC.

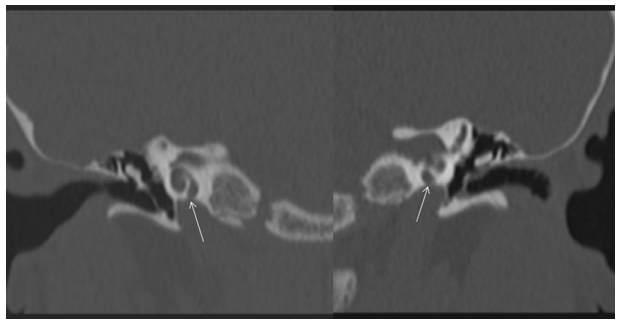

Figure 8 Carotid- Cochlear Dehiscence. On Coronal images of a MSCT of the temporal bone, bilateral dehiscence of the cochlea is noted into the carotid artery (arrow).

Otosclerosis is a disorder of the bony labyrinth characterized by an active metaplastic process within the endochondral layer of the temporal bone. Cavitating otosclerosis results in cavity formation within an otosclerotic focus that is continuous with CSF.[43] The pathologic third window communication is presumed on the scala vestibuli site of the endosteum of the cochlea or vestibule[44] (Fig. 9).

In Paget's disease, there is diffuse involvement of the otic capsule. On histologic examination, multiple microfractures within the otic capsule on the scala vestibuli side of the cochlear partition and extensive Pagetic involvement of the otic capsule surrounding the labyrinth are noted. It is postulated that the microfractures and/or Pagetic bone act as a distributed third window, dissipating sound energy transmitted through the stapes footplate away from the cochlea.[3,29]

In summary, mobile or pathologic third window lesions present with classic signs and symptoms. CHL can mimic middle ear disease clinically. On audiogram, there is a low frequency air bone gap. The lesions reside on the scala vestibuli side of the cochlear partition. Lesions may be focal and may involve the semicircular canal, vestibule, cochlea, or lesions may be diffuse. Thin collimation multislice CT is essential to make an accurate radiologic diagnosis.

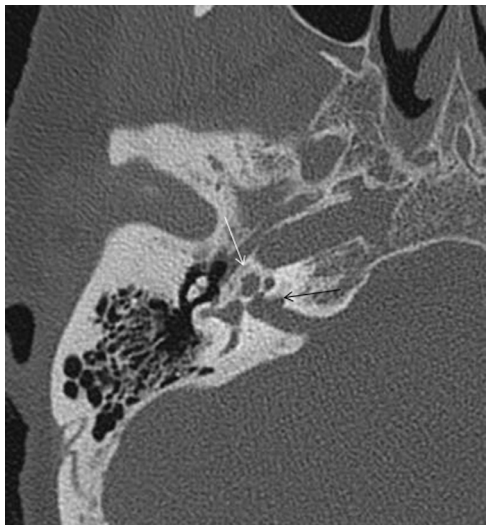

Figure 9 Cavitating Otosclerosis. Axial MSCT of the temporal bone demonstrates pericochlear lucency (white arrow) compatible with otosclerosis. There is also a CSF density diverticulum arising from the IAC (black arrow) representing cavitating otosclerosis.

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