Chapter 6
Third Mobile Window Syndromes

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Learning Objectives

- The normal inner ear has openings at the round and oval window. Additional or third windows can be the source of symptoms including conductive hearing loss, dizziness, oscillopsia, autophony, and bone conducted hyperacusis.
- Pathophysiology of third window syndromes.
- Different types of third window syndromes have been described.
- Symptoms that are suggestive of a third window syndrome, and other possible causes of these symptoms that should be considered in the differential diagnosis.
- Appropriate work up, and limitations of testing.
- Available treatment options including risks and potential benefits.

Introduction

The normal inner ear has two mobile windows which are not surrounded by fixed bone. These are the oval window where the stapes interfaces with the inner ear and the round window. In a normal inner ear, any displacement of the stapes results in a pressure wave that travels through the cochlea to the round window. Third mobile window syndromes represent a pathological extra opening into the inner ear. The best known and most extensively studied of these superior canal dehiscence

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B. T. Crane et al. (eds.), *Disorders of the Vestibular System*,
https://doi.org/10.1007/978-3-031-40524-2_6

syndrome (SCDS) [1] has well understood pathophysiology, and curative surgery therapy is now regularly performed throughout the world [2]. Of course it is also possible to have a pathologic opening in almost any other area of the inner ear including other semicircular canals [3, 4], or the cochlea such as near the facial nerve [5, 6]. The pathophysiology of these conditions is due to this "third mobile window" in addition to the oval and round windows [7]. With an abnormal opening in the labyrinth, pressure entering the labyrinth via the stapes at the oval window is partially shunted away from the round window to a new low impedance canal pathway in the canal. Bone conducted sound also now has a new route of entrance into the inner ear via the dehiscence.

Posterior semicircular canal dehiscence is now recognized as a third mobile window syndrome [3, 8]. This condition usually also presents with pulsatile tinnitus, sound or pressure induced vertigo, and bone conduction hyperacusis symptoms. Other atypical symptoms which can mimic Meniere's disease have been described [9, 10]. Dehiscence of the posterior canal can be associated with jugular bulb anomalies, fibrous dysplasia [11], congenital syndromes, cholesteatoma [12], and iatrogenic injury. In a recent review, the most common presenting symptoms were sound induced vertigo (38%), mixed hearing loss (36%), and tinnitus (34%) [3]. Thus, symptoms are often similar to SCDS, although like SCDS, it is believed that many patients may be asymptomatic and never present for evaluation. Transmastoid plugging of the posterior canal seems to be the most the most common treatment for this condition [8, 12, 13].

Lateral semicircular canal dehiscence usually occurs as a complication of chronic otitis media [14], cholesteatoma, and associated surgical treatment. Such fistulas are found in 2% of revision canal wall up mastoidectomy and 13% of revision canal wall down mastoidectomy [15]. Management of these fistulas is controversial some have advocated a complete removal of the cholesteatoma with repair of the bony defect [16], while others have advocated leave some cholesteatoma over the fistula and doing a canal wall down procedure [17].

Enlarged vestibular aqueduct syndrome (EVAS) is also recognized as a cause of third mobile window syndrome due to a larger than normal vestibular acting as a path for acoustic energy to be shunted away from the cochlea. The criteria used for diagnosis is typically the midpoint of the vestibular aqueduct being 1.5 mm or greater [18]. The finding is often bilateral and can be associated with other conditions such as Pendred syndrome and choanal atresia. Hearing loss is often the predominate feature and it is often conductive although a sensorneural hearing loss is also present in many cases. Perhaps because acoustic energy is not shunted through the semicircular canals, only rarely is vertigo a symptom of EVAS [19]. Surgical treatment associated with closing or reducing the size of the vestibular aqueduct has not been successful.

Discussion will now focus on SCDS as this is the most common and most extensively studied third window syndrome, and because other third window syndromes often include similar symptoms. Patients with SCDS can present clinically with sound-induced vertigo and oscillopsia (bouncing of vision), and with decreased hearing thresholds for bone conducted sounds (i.e., conductive hyperacusis) while having increased thresholds for air conducted sounds. This can lead to difficulty

hearing external air conducted sound but also causes heighted perception of bone conducted sound so patients can hear their pulse, eye movements, chewing or steps as well as experience autophony. The dizziness and vertigo symptoms are common and can be disabling. These symptoms include chronic disequilibrium as well as vertigo induced by loud sound or pressure changes [2, 20, 21].

Diagnostic Evaluation

As with any complaint of dizziness, a good history is key. Patients with SCDS usually present with a primary complaint of either autophony or dizziness although they occasionally have isolated conductive hearing loss [22] that can mimic otosclerosis. Vertigo related to SCDS is usually brief and induced by loud sound or pressure changes. Sound-induced dizziness or oscillopsia is present in 90% of SCDS patients [21]. Pressure induced vestibular symptoms, often manifest with coughing or straining, are present in 73% of patients, with 67% exhibiting both pressure- and sound-related symptoms [21]. Chronic disequilibrium may also be attributed to SCDS [23].

Auditory symptoms are common and may be present in 85% of SCDS cases [24]. Hyperacusis for bone-conducted sound [25, 26] is present in 52% [21]. Symptoms often include hearing one's pulse or eye movements. Autophony or the patients hearing their own voice sounding disturbing to them is present in up to 60% of patients [21, 27].

Eye movements in the plane of the superior canal evoked by sound or pressure are the hallmark clinical finding of SCDS [28, 29]. The eyes should be examined under Frenzel or video goggles to eliminate visual fixation. Tones at levels up to 110 dB nHL should be delivered in one ear at a time. Sound-evoked eye movements were noted in 82% of SCDS patients [21]. Eye movements can also be induced with Valsalva maneuvers (75%) or pressure applied to the external auditory canal (45%). Depending on the type of stimulus and the direction of endolymph flow either excitation or inhibition of the superior canal may occur. Eye movements evoked by pressure or sound almost always occur in the superior canal plane. If eye movements are in another direction, the SCDS diagnosis should be questioned, and alternative diagnoses of posterior canal dehiscence [30] or horizontal canal fistula [31] considered. Sound-evoked rotation of the head in the superior canal plane occurred in 14% of patients.

The audiogram is an important part of the SCDS evaluation, and a minority of patients have only auditory symptoms [21, 22, 32, 33]. Conductive hearing loss (CHL) is often largest at lower frequencies [22, 32, 34], and bone conduction thresholds can be negative (conductive hyperacusis). Because of the CHL and normal appearance of the ear, some patients with primarily auditory symptoms have been misdiagnosed as having otosclerosis [33]. The key differences are (1) that conductive *hyperacusis* does not occur in otosclerosis, and (2) that the acoustic stapedial reflex, which is often normal in superior canal dehiscence, should be absent with otosclerosis.

Vestibular-evoked myogenic potentials (VEMP) responses are enhanced in SCDS. The cervical VEMP (cVEMP) is measured from the sternocleidomastoid muscles using averaged electromyography in response to multiple loud clicks or tone bursts delivered to the ear. The reflex is thought to be activated by sound transmitted through the stapes footplate to the saccule which is innervated by the inferior vestibular nerve [35]. Decreased cVEMP thresholds and large amplitude responses are indicative of SCDS. cVEMP thresholds for air-conducted 500 Hz tone bursts, for example, cVEMP thresholds were 80–95 dB SPL for 13 patients with SCDS (83.85 ± 1.40 dB SPL, mean ± SD), 20–30 dB lower than in normal control subjects (110.25 ± 1.28 dB SPL) [36]. It has been argued that cVEMP is better than 90% sensitive and specific for SCD [37] but sensitivity and specificity depend on the parameters used [38]. Ocular VEMP (oVEMP) similarly measures averaged electromyography in response to tone bursts delivered to the ear or sometimes forehead taps. The oVEMP reflex is thought to be activated by sound or vibration being transmitted via the utricle and superior vestibular nerve to the contralateral inferior oblique muscle [39, 40]. The amplitude of this excitatory potential is measured through surface recording electrodes placed beneath the eyes. Some prefer the evaluation oVEMP responses as they may be more easily tolerated and more sensitive and specific for detection of SCDS than the cVEMP [41–43]. The oVEMP responses can also be done more quickly as they do not require finding a threshold. As a result, oVEMP testing is becoming more widely available at academic centers. Despite its utility for diagnosis, these tests are poorly correlated with patient symptom severity [44].

The VEMP is not always measurable and is likely to be absent in those with previous middle ear surgery. Although the oVEMP measurements can circumvent this problem using forehead taps [42], this version of the test is less commonly available. The VEMP threshold may also be decreased in other conditions such as enlarged vestibular aqueduct syndrome [45]. Use of VEMP can also differentiate SCDS from middle ear causes of conductive hearing loss in which VEMP should be absent [46].

Imaging of the temporal bone using computed tomography (CT) must show the absence of bone over the superior canal (SC) for SCDS to be considered. If the SC appears covered by bone on CT the diagnosis of SCDS is excluded; however, the appearance of a dehiscence on CT does not rule out thin bone covering the SC below the resolution of the scanner. Thus, CT is a highly sensitive test for SCD but it is not specific [47] due to a high rate of false positives. In a review of temporal bone CT scans done in the general population, 9% of scans had apparent SCD with one observer calling as many as 12% [48]. Many are likely false positives caused by the limits of resolving thin bone, since the incidence of SCD in a survey of temporal bones was only 0.7% [49]. Images should be reconstructed in the plane of the superior canal as well as orthogonal to it so that any dehiscence can be definitively demonstrated. However, due to the high risk of false-positive findings and overestimation of dehiscence size [50], the diagnosis of SCD must never be based on a CT alone.

Magnetic resonance imaging (MRI) may suggest SCD [51]. The best images are a T2-weighted protocols (e.g., Constructive Interference in Steady State, or CISS) and reconstructed in the plane of the superior canal. However, even these protocols

Table 6.1 Proposed diagnostic criteria for superior semicircular canal dehiscence syndrome (SCDS)

Diagnostic criteria for superior semicircular canal dehiscence syndrome
1. At least one of the following symptoms consistent with a third window lesion of the inner ear:
(a) Bone conduction hyperacusis
(b) Pulsatile tinnitus
(c) Acute sound-induced vertigo and/or oscillopsia
(d) Pressure-induced vertigo and/or oscillopsia
2. At least one of the following diagnostic tests indicating a third mobile window of the inner ear:
(a) Nystagmus characteristic of excitation or inhibition of the affected superior canal evoked by sound (Tullio phenomenon) or changes in middle ear pressure (Hennebert sign) or intracranial pressure
(b) Low-frequency negative bone conduction thresholds on pure tone audiometry
(c) Enhanced VEMP responses (low cervical VEMP thresholds or high ocular VEMP amplitudes)
3. High-resolution temporal bone CT imaging with multiplanar reconstructions demonstrating dehiscence of the superior canal
4. Not better accounted for by another vestibular disease or disorder

may have a high false-positive rate, and CT is probably the better study for most evaluations. However, MRI may be appropriate for evaluating the efficacy of previous canal plugging by assessing the fluid signal in the superior canal [52].

The diagnosis of SCDS should be based on CT imaging showing a dehiscence, symptoms consistent with a mobile third window, and at least one physiologic measure supporting the presence of a third mobile window. The proposed diagnostic criteria for diagnosis of SCDS [2] are shown in Table 6.1.

Differential Diagnosis

Other common conditions can cause symptoms similar to SCDS making it important to consider a differential diagnosis. When conductive hearing loss is present in a setting without trauma and with a normal otoscopic exam, SCDS should be considered along with otosclerosis. Autophony raises the possibility of a patulous Eustachian tube, but SCDS can produce a similar sensation. Episodic vertigo evoked by intracranial or middle ear pressure changes could indicate a perilymphatic fistula, but SCDS should strongly be considered as an alternative diagnosis. We have seen several patients who have undergone previous surgical explorations for these presumed otological disorders, only later to be found to have SCDS.

The conductive hearing loss with SCDS often appears similar to otosclerosis because both occur in adulthood with a normal otoscopic exam [33]. The audiograms differ in that patients with SCDS often have negative bone conduction thresholds, and if there is no previous history of middle ear surgery the acoustic reflex is often intact.

Autophony is often the predominant symptom in patients with a patulous Eustachian tube (PET) [53], but it can also be the most disturbing symptom in

SCDS [54]. One distinguishing feature is that patients with PET typically have autophony for their breath sounds (especially for nasal breathing), whereas patients with SCDS usually do not [53]. A history of vertigo and hyperacusis to bone conducted sound are atypical of a PET. The audiogram, VEMP, and CT will typically differentiate a PET from SCDS.

Perilymph fistula and fenestrations of other semicircular canals are considered in the differential diagnosis of SCDS [4, 55, 56]. The diagnosis of perilymph fistula is most clear in the presence of recent stapes surgery, temporal bone fracture, or barotrauma injury. In these cases, acute vertigo is usually accompanied by a sensorineural hearing loss. A fistula in the horizontal canal can be acquired in cases of cholesteatoma or prior mastoidectomy [15]. Spontaneous perilymph fistula is a controversial diagnosis, which if considered at all should only be considered after all other possible causes are excluded [57].

The most common cause of spontaneous (nonpositional) vertigo is vestibular migraine [58–60]. The migraine incidence is 17.6% of females and 5.7% of males [61], and approximately 25% of these report associated vertigo [62]. Migraine is much more common than SCDS, and inevitably symptoms in some patients with radiographically apparent SCD with nonspecific symptoms may be better explained by migraine. Particularly challenging are those patients who have both SCDS and migraine [2, 63]. It may be difficult to determine if their sound sensitivity is due to one more than the other, for example. Their chronic disequilibrium may be related to migraine, or it may be due to the constant transmission of intracranial pressure pulsations through the dehiscence. The symptoms of SCDS could serve as triggers to exacerbate migraine in susceptible individuals. However, the neurotologist must also consider that failure to recognize and treat coexistent migraine can lead to disappointing results in SCDS surgery, as it can with other causes of vertigo. Optimization of migraine treatment is recommended prior to SCD surgery [2, 63].

Operative Decision Making

The physician and patient must weigh the symptom severity against the risks and benefits of surgery. In the authors' experience, only a third of patients with SCDS elect to have surgery, with the remaining patients choosing to live with their symptoms or making lifestyle changes to avoid situations that exacerbate symptoms. Control of comorbid vestibular migraine has in several cases allowed patients to avoid surgery.

Most SCDS patients present to a neurotology clinic for dizziness or vertigo of variable severity. Some patients are disabled by their symptoms, and surgery is the only viable option for them to have an acceptable quality of life.

Autophony, or the abnormal sound of one's own voice, can be disabling. There is no medical treatment for autophony symptoms due to SCDS, as the sound transmission is via bone, not the Eustachian tube. Thus, for SCDS patients who are

significantly disturbed by autophony, surgery is the only option for relief and has been shown to have a significant benefit [54, 64].

Conductive hearing loss is common in SCDS [22, 27, 65, 66], but it is often limited to low frequencies and one ear, so many patients do not have a significant disability from it. The risk of hearing loss progressing over time is also low [67, 68]. In most patients, the conductive hearing loss improves with surgery [65, 69]. However, plugging of SCD also carries a risk of hearing loss [70], which is greater in patients with prior ear surgery [65, 69]. Patients who have hearing loss as their primary symptom of SCDS should be encouraged to consider nonsurgical options such as a hearing aid.

Pulsatile tinnitus and bone conduction hyperacusis can be disabling in some and the primary reason to seek treatment. In rare cases, nonsurgical options can ameliorate these symptoms for instance in cases were the SCDS occurs at the superior petrosal sinus, embolization [71], or stenting [72] of the superior petrosal sinus has improved symptoms.

Although dizziness symptoms are often the motivation for surgery, imbalance symptoms may be worse during the immediate postoperative period. Symptoms improve as the patient adapts, and we typically prescribe vestibular rehabilitation. Benign paroxysmal positional vertigo (BPPV) has been reported in as many as 24% of cases [73] after plugging, and can be treated with repositioning maneuvers. Plugging of the superior canal will cause loss of function due to hydrodynamic insufficiency of the plugged canal [74, 75]. However, patients can adapt very well to this single-canal insufficiency, as low-frequency, low-acceleration head movements still generate useful inhibitory signals from the contralateral posterior canal.

Bilateral Dehiscence

About a quarter of individuals with SCDS have the appearance of bilateral SCD on high-resolution CT scan [76]. Fortunately, one side is usually responsible for most symptoms. In some cases, symptoms and signs can be elicited from both ears. In such patients that do have bilateral SCDS, every effort should be made to identify the more symptomatic ear and operate on that side first. In most cases, symptoms will either resolve after operating on the more symptomatic side or abate to the point that contralateral surgery is not required. Only 11% of patients with bilateral SCDS opt to have bilateral surgery [76]. We recommend the second side only be considered for plugging surgery after sufficient time for adaptation in the partial loss of vertical semicircular canal function, typically after 6 or more months have passed since the operation. Plugging of both superior canals significantly impairs the ability to sense downward head rotation in the vertical plane, so these patients are at risk of developing vertical oscillopsia during ambulation [76]. Patients with bilateral SCDS had worse symptom control than those with unilateral disease if one or both sides were treated [64].

Near Dehiscence

Although controversial, it is recognized that symptoms of SCDS can occur even if very thin bone over remains over the superior canal [77]. In these cases, CT is often read as showing a dehiscence, and the patient can have other objective evidence of dehiscence including an air-bone gap on audiometry, increased oVEMP amplitude, and low cVEMP thresholds. Although these patients can benefit from surgery, caution is suggested as these patients have more residual symptoms and may have a higher rate of postoperative complications than patients with frank dehiscence [2] but they can, in some cases, benefit from surgery [78].

Operative Technique

The middle cranial fossa approach was described first [1] and is the technique detailed in the following paragraphs. An alternative transmastoid approach has also become popular. Advocates of the transmastoid approach have noted that it avoids a craniotomy, involves no temporal lobe retraction, and may lead to better stability of the canal plug. Moreover, most otolaryngologists are more familiar with mastoidectomy [79, 80]. The transmastoid approach was initially described in two patients in 2001, and although these patients were relieved of vertigo symptoms, one patient experienced significant sensorineural hearing loss after surgery [81]. More recently additional reports of transmastoid superior canal plugging have been published with both minimal morbidity and improvement in symptoms [2, 79, 80, 82–86].

The middle fossa approach has, in principle, some advantages. The transmastoid approach does not allow direct confirmation of the dehiscence, and transmastoid plugging of a superior canal that was later found to be intact has been described [79]. The transmastoid approach may not be possible in patients with a low hanging dura or extensive tegmen dehiscences [79]. In the transmastoid approach, openings in the canal need to be created and plugging material must be advanced beyond these openings to be successful. Thus, the plug is placed closer to the sensory epithelia of the ampulla and the utricle. This may be more traumatic to these structures, risking disturbance of their baseline firing rates. Furthermore, opening the superior canal distal to the dehiscence may place the plug into the common crus, causing loss of sensory function of the posterior canal as well [75]. The transmastoid approach also creates a new dehiscence which can make it difficult to know the source if the patient develops residual symptoms later. Finally, the transmastoid approach requires drilling, irrigation, and suctioning on the bony canal. Once the canal is opened, these manipulations could contaminate or remove perilymph from the canal and cause collapse of the membranous labyrinth or serous labyrinthitis.

Round window plugging or reinforcement has been suggested as a treatment for SCDS [87, 88]. The procedure does not directly address the third window at the superior canal and some patients developed worse symptoms after this procedure

[87]. A recent retrospective case review suggested that although some patients reported improvement in subjective symptoms such as autophony and vertigo, improvement in objective tests such as VEMP were rare and hearing was often diminished after the procedure [89]. Many of these patients require revision surgery via a transmastoid or middle fossa approach [90]. Due to the poor outcomes and because it does not directly address the known pathophysiology of the disease, round window plugging is not widely considered to be an appropriate treatment [89, 91].

The transmastoid approach may be preferable in cases where the patient cannot tolerate a middle fossa surgery, or the dehiscence cannot easily be accessed through the middle fossa—for instance when the dehiscence is at the superior petrosal sinus [92, 93]. Some patients who have been symptomatic after a middle fossa approach have had relief of symptoms with a revision surgery via a transmastoid approach [83]. It is difficult to directly compare outcomes associated with a transmastoid and middle fossa approach because of differences in relative indications between them.

For middle fossa approach, the incision is made from the helical root around the helix to a location over the external auditory canal, and then superiorly. Temporalis fascia is harvested for later use in plugging the superior canal and for repair of any tegmen defects or cerebrospinal fluid leak that may occur. Afterward, the temporalis muscle is divided, and the area of the craniotomy exposed.

The craniotomy should be centered over the superior canal, the external auditory canal is often a good landmark but image navigation can be used. The lower border of the craniotomy is placed just high enough to avoid the mastoid air cells. The width and height of the craniotomy should accommodate a middle fossa retractor, typically 3 cm wide by 4 cm high. The craniotomy is opened by drilling troughs around the borders using a 4-mm burr. The dura should remain intact, and the bone flap removed and preserved in saline. The dura is further elevated from the edges of the craniotomy to allow retraction. The sharp edges of the craniotomy are removed using small Kerrison rongeurs, and the bone chips created in this process can be used as plugs for the superior canal.

The middle cranial fossa retractor is placed and used to gently elevate the dura off the middle fossa. Dura here can be thin, especially if tegmen dehiscences are also present, and large cotton balls soaked in saline are a minimally traumatic means for the dural elevation. A hemostatic agent such as dry microfibrillar collagen (Avitene®) or gelatin powder (Gelfoam®) mixed as a paste with thrombin is generously applied in advance of the cotton balls. The surgeon is careful to only suction on the cotton balls and not to directly suction the area of the dehiscence because of the risk that this poses for removing excessive perilymph or for tearing the membranous labyrinth, which could cause sensorineural hearing and vestibular loss.

After identifying the superior canal dehiscence, attention is shifted toward plugging. Small pieces of previously harvested temporalis fascia are gently slid into the two open lumens of the bony superior canal. Several pieces are used to advance the plugs a few millimeters beyond the dehiscence. Care must be taken that one end is not thus left open because its fascia is displaced, which is common as fascia is pushed in the other side. To prevent this, once the fascia is in place, bone chips

matching the diameter of the canal are firmly lodged so as to "cork" each end of the dehiscence. Other groups have used materials such as bone wax [94] or a mixture of fibrin glue and bone dust [79]. The surgeon must ensure a watertight seal is obtained to prevent pressure transmission through the third mobile window. Bone cement can also be used to resurface the area after plugging.

Closure is achieved by anchoring the previously harvested bone flap in place. The temporalis muscle is reapproximated with absorbable sutures, and the skin is closed with staples and/or suture. A drain is not typically used, but a gentle pressure dressing is maintained for 2 days.

Postoperative Care

A monitored bed with neurological checks in the immediate postoperative period is recommended due to the epidural hematoma risk. Postoperative patients are treated with intravenous steroids which can be quickly tapered. Patients frequently experience nausea during the initial hours after surgery. This is best controlled with intravenous promethazine (Phenergan). For the first 24–26 h, short acting narcotics can be administered by the patient-controlled analgesia (PCA) with proper neurological nursing assessments to ensure that any change in neurological status is not masked by excessive sedation. Routine postoperative analgesics are sufficient to control the pain thereafter. If the patient is experiencing intense pain or if there is any change in mental status, an epidural hematoma may be the cause and an immediate head CT should be considered. The typical hospitalization lasts a total of 2 or 3 days.

Long-Term Results

Most patients are extremely satisfied with the surgery, with studies supporting improvements in overall quality of life [86, 95], autophony [54], and dizziness symptoms [96]. Relief of dizzy symptoms has been documented by measuring the dizziness handicap inventory (DHI) [97] which improved by 26 points. Patients with more severe dizziness (DHI $\geq$30) improving by an average of 39 points [96]. Nearly all patients would recommend the surgery to others [64].

For some patients, autophony or hyperacusis for internal sounds are the primary reason for undergoing SCDS surgery and this is the most reported presenting symptom after dizziness [27]. Autophony is on average 89% improved immediately after surgery [54] and similar improvement is maintained long term [64]. Some autophony symptoms may take time to resolve due to fluid collecting in the middle ear after surgery.

The results for improving hearing with SCD surgery are gratifying if conductive hyperacusis is documented preoperatively. Dramatic results have been reported in some patients, [98] but are uncommon. The air-bone gap that is present prior to surgery typically closes within several months after surgery [69, 99] once any

middle ear effusions or hemotympanum have resolved; however, patients can also experience hearing loss after SCD surgery. Two larger series found a mild (~10 db) high-frequency sensorineural hearing loss in 25% of cases [69, 99] and profound hearing loss has been reported in 2.5% of cases [100]. In patients with previous middle cranial fossa or stapes surgery, one series found the risk of hearing loss was high [65]. A recent review found audiometric outcomes varied significantly among studies and although transmastoid and middle fossa approaches seem to be safe, subjective hearing improvement was not significant [101]. However, our own experience is that air-bone gaps, if present prior to surgery, are reduced after surgical plugging of the affected canal, and that symptoms of conductive hyperacusis like autophony and pulsatile tinnitus are also ameliorated.

Summary

The diagnosis of SCDS is based on patient history, physical exam including eye movements in response to sound or pressure, and other supporting studies including the audiogram, VEMPs, and CT imaging. The spectrum and severity of symptoms of SCDS vary significantly among individuals, and the potential benefit of surgery must be carefully compared to the risks and probability of success in each patient. A large fraction of patients with SCDS do not opt for surgery.

Both middle fossa and transmastoid approaches are reasonable treatments for SCDS. Patients generally experience an improvement in symptoms of dizziness, autophony, and hyperacusis symptoms. Although there is often an improvement in hearing after surgery, this must be carefully weighed against the risk of hearing loss, which is significant in patients who have had previous middle fossa or stapes surgery.

Quiz Questions

1. True/False: Superior canal dehiscence is usually congenital.
2. True/False: Horizontal canal dehiscence is occurs as a complication of cholesteatoma or mastoidectomy surgery.
3. True/False: Computed tomography (CT) is the gold standard for diagnosis of SCD, and if it is seen on CT, no further testing is required for diagnosis.
4. True/False: Vestibular migraine is much more common than third window symptoms and should be treated prior to considering treatment for SCDS.
5. True/False: When the threshold of cervical vestibular-evoked myopotentials is higher than 95 dB nHL, it suggests SCDS.
6. True/False: Ocular vestibular-evoked myopotentials have a larger than normal amplitude in patients with SCDS.
7. True/False: Transmastoid plugging of the superior canal is a good option for patients who are not candidates for a middle fossa approach.
8. True/False: Plugging the round window directly addresses the site of the dehiscence in third window syndromes.

9. True/False: The majority of patients with SCDS opt to get surgical treatment.
10. True/False: Conductive hearing loss with an absent acoustic reflex and otherwise normal ear exam suggests SCDS.

Quiz Answers

1. False. Congenital SCD is very uncommon.
2. True. The horizontal canal is the most frequent site of violation of the inner ear due to cholesteatoma and related surgery.
3. False. CT has a high positive rate with about 10% of scans showing dehiscence, while the true incidence is probably closer to 1%.
4. True. Vestibular migraine is much more common than SCDS and other third window syndromes.
5. False. Threshold of cVEMP above 95 dB nHL is normal. cVEMP thresholds in SCDS are typically lower than normal. The exact threshold may depend on the lab and technique but typically less than 75 dB.
6. True. Large oVEMP amplitudes suggest SCDS.
7. True. Most agree either the transmastoid or middle fossa approaches are reasonable.
8. False. The round window is part of normal inner ear physiology, a third window must occur at other site. Round window plugging is not considered to be standard of care.
9. False. In several series, about one in three patients opts to get surgery. Many patients with superior canal dehiscence probably never develop symptoms.
10. False: Conductive hearing loss with an absent acoustic reflex suggests otosclerosis. In SCDS, the acoustic reflex should be present.

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