Chapter 5
The Otologic Mimicker: Vestibular and Auditory Symptoms

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Introduction

It is said that third mobile window disorders (TMWD) are the great otologic mimicker, presenting with vestibular and auditory symptoms mimicking some of the more common otologic disorders making the accurate diagnosis of a TMWD all the more difficult. This chapter will help differentiate the classic symptoms of common and less common otologic disorders to help the clinician make accurate diagnoses. We will discuss the classic findings and symptoms found in a patient with a TMWD and the testing that will assist in ruling out other disorders to make the correct diagnosis.

Vestibular Symptoms

Without exception, history taking is the single most important diagnostic tool for a patient that presents with vertigo or dizziness. An accurate diagnosis can be obtained 80% of the time based on history alone [1, 2]. Some of the important aspects of the

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history to ascertain are description of events, duration, frequency, triggers, changes in hearing, exacerbating factors, and what resolves the events. True vertigo is generally broken down into central vs. peripheral. Other common etiologies can cause vertigo-like symptoms including cardiac, neurologic, metabolic disorders, or medication side effects that patients may perceive as vertigo. Physical exam and further vestibular testing can help elucidate the true underlying etiology of a patient's vertigo.

Subjective Findings

Possibly the most integral part of a patient's history is the description of their "dizzy" episodes. Dizziness can have varying meanings to different people. Is it a true room spinning vertigo with a sensation of movement indicating a likely peripheral etiology? Or is it a disequilibrium, "feels like my balance is off," that may suggest a central etiology? Central disorders classically include retrocochlear or cerebellar dysfunction/lesions. Peripheral vertigo can be seen in numerous conditions including unilateral vestibular weakness, benign paroxysmal positional vertigo (BPPV), Mal de Débarquement syndrome, Ménière's Disease, or TMWD. Patients describing lightheadedness/presyncope or vague "mental fog" suggests a non-vestibular disorder and more of a systemic etiology including arrhythmias, cardiogenic, anemia or poor circulation, neurogenic, thyroid issues, orthostatic hypotension, etc. A thorough non-vestibular workup should be performed in these patients by their primary care provider. In the patient that describes rotary vertigo, their description of the events should include queries regarding duration, frequency, and associated symptoms such as changes in hearing, aural fullness, and headaches. Triggering or exacerbating factors are particularly important to explicate such as movement induced symptoms, noise induced symptoms, Valsalva triggers, recent upper respiratory infections, stress, high-salt diet, allergies, barotrauma, or weather changes. Equally important is what helps to abort the episodes: eye fixation, going to a dark quiet room, or medications.

TMWD represent areas of dehiscence of the bony labyrinth or inner ear (including the round and oval windows) that creates a characteristic vertigo triggered when the area of dehiscence is subjected to pressure change. External pressure can be presented in different forms. Sound-induced vertigo is the classically described Tullio phenomenon while pressure-induced vertigo from pneumatic otoscopy describes the Hennebert sign or "fistula sign." Both of these signs result in a vertical nystagmus first described by Minor and colleagues in 1998 with respect to superior semicircular canal dehiscence (SSCD) [3]. Tullio phenomenon was described by an Italian biologist, Pietro Tullio, in 1929 when he discovered that a fistula created in the horizontal semicircular canal of pigeons resulted in the birds quickly turning their heads in the contralateral direction when exposed to loud sounds [4]. This first became clinically relevant when Hennebert made the connection between pressure-induced vestibular changes and inner ear dysfunction in patients with congenital syphilis. Later temporal bones of syphilitic patients were found to have gummatous

osteomyelitis and fistulas of the labyrinth [5]. Vertigo in TMWDs with meningeal exposure, as occurs in SSCD, can also be triggered by a Valsalva maneuver or any acute change in intracranial pressure. Provocative or exacerbating factors of a patient's vertigo are particular symptoms that may clue a provider into a possible TMWD diagnosis.

Vestibular symptoms from SSCD were divided into four categories by Minor with associated prevalence [6]:

- Tullio phenomenon, eye movement evoked by sound: 82%.
- Valsalva-induced, eye movement evoked by internal pressure: 75%.
- Hennebert sign, eye movement evoked by external pressure on the tympanic membrane: 45%.
- Sound-induced head tilt in the plane of the affected canal: 20%.

In addition to episodic vertigo, chronic disequilibrium is a common complaint of patients suffering from a TMWD—affecting up to 76% in one case series [7]. The disequilibrium may or may not worsen with sound or external pressure. Patients often have a difficult time describing their vestibular symptoms, which can be quite debilitating. The wide variety of patient descriptions of vestibular symptoms in TMWD is one of the reasons it is called the otologic mimicker.

Physical Exam Findings

Vestibular examination can help to differentiate TMWD from other common vestibular disorders. Most patients with small to moderately sized TMWD will demonstrate normal and symmetric vestibulo-ocular reflexes on head thrust testing and the absence of nystagmus after horizontal or vertical head shaking. Defects greater than or equal to 5 mm in SSCD will start to show nystagmus on head thrust testing [3]. Spontaneous nystagmus is not typically seen in TMWD although it has been described in rare cases due to large defects in the superior semicircular canal (SSC) that allow intracranial pressure variations to create a pulsatile stimulus [8].

Findings characteristic of a peripheral vestibular origin include spontaneous nystagmus with the head still, decreased nystagmus with visual fixation, and/or increased nystagmus when fixation is absent [9]. Infrared video goggles or Frenzel glasses can help facilitate testing, allowing the practitioner to better assess nystagmus characteristics by preventing visual fixation. Misalignment of the eyes, i.e., strabismus, while not a vestibular disorder can certainly produce symptoms of vertigo/dizziness and may be apparent with use of Frenzel lenses. Ophthalmology referral is indicated for such patients.

A Dix-Hallpike test should be performed on all patients presenting with vestibular symptoms to assess for possible BPPV, the most common cause of peripheral vertigo. Testing should be performed even if symptoms appear non-positional. The Dix-Hallpike maneuver is performed by rotating the patient's head 30–45° towards the ear being tested, starting in the sitting position and quickly placing the patient in

Table 5.1 Vestibular mimickers

Signs/symptoms of TWS	Differential diagnosis
Vertigo/dizziness	BPPV; migraine; Ménière's disease; labyrinthitis; AIED; mass lesions (vestibular schwannoma); TWS; Mal de Débarquement; central etiology (stroke/TIA)
Tullio's phenomenon (vertigo with loud sounds)	Ménière's disease; otosyphilis; TWS; idiopathic; vestibulocochlear fibrosis; postsurgical; lyme disease; otosclerosis
Hennebert's sign (pressure induced vertigo through the EAC)	Ménière's disease; otosyphilis; TWS
Visual-spacial disorientation	Migraine; multiple sclerosis; multisensory balance dysfunction; cognitive dysfunction (Alzheimer's disease)
Valsalva induced vertigo	Vertebrobasilar insufficiency; TWS

the supine position. A positive test will evoke a geotropic rotary nystagmus indicating otolith presence in the testing ear's posterior semicircular canal. The test can conveniently be transitioned into a canalith repositioning maneuver (i.e., Epley maneuver) to reposition the otoliths out of the posterior SCC.

Table 5.1 outlines common vestibular symptoms associated with TMWD and a possible differential diagnosis for each symptom. Exploration of diagnostic vestibular testing related to TMWD can be explored in Chap. 11.

Auditory Symptoms

As in the patient presenting with vestibular complaints, a thorough history is the centerpiece to working up a patient presenting with auditory symptoms. The presence of otalgia, otorrhea, tinnitus, aural fullness, hearing loss, and fluctuation of hearing must all be explored in the patient interview. The duration and frequency of symptoms, a history of ear infections or prior ear surgeries, exposure to ototoxic medications or loud noise, and a history of head trauma are additional and essential aspects of the history to obtain. A tuning fork exam can be performed in the office, but ultimately a full audiogram should be performed.

Subjective Findings

Auditory findings in TMWD, like vestibular findings, can vary widely. Common symptoms often described by TMWD patients include autophony, hearing internal bodily movements, aural fullness, hearing loss, hyperacusis, and pulsatile tinnitus. In addition to hearing one's voice, some patients may describe being able to hear

their eyeballs move or their feet hit the floor; this description differs from the autophony observed in patulous eustachian tube dysfunction (ETD) that is exacerbated by respiration and correlates with coordinated tympanic membrane movement on exam. Patients with negative pressure ETD sometimes also describe autophony but this is differentiated from TMWD by retracted tympanic membrane or middle ear effusion on physical exam and abnormal tympanogram findings. Aural fullness is a common complaint in a number of disorders including low frequency sensorineural hearing loss (LFSNHL), Ménière's disease, ETD, and temporomandibular myofascial disorders. TMWDs with either meningeal or vascular bony dehiscence can both present with pulsatile tinnitus, which can also present in vascular lesions (arteriovenous malformation (AVM)/arteriovenous fistula (AVF)), idiopathic intracranial hypertension (IIH), glomus tumors, venous hum, sigmoid sinus diverticulum, and carotid pseudoaneurysm. Appropriate imaging modalities will help to differentiate these underlying etiologies. Conductive hyperacusis including a feeling or hearing the pulse in the affected ear, has been described in up to 39% of patients with SSCD [10]. Ultimately, it is less common for TMWD to present with auditory symptoms alone without any coexisting vestibular complaints.

Physical Exam Findings

Patients with TMWD will classically demonstrate a conductive hearing loss on tuning fork examination with a 512 Hz Weber test lateralizing to the ipsilateral ear [3, 11]. 54% of patients will demonstrate a vertical–torsional nystagmus on pneumatic otoscopy [10]. On otoscopy, in an absence of a history of ear disease, the external auditory canal, tympanic membrane, and mesotympanic space will appear normal. An abnormal otoscopic examination should lead the practitioner down the diagnostic pathway for the visualized lesion.

Table 5.2 outlines common audiologic symptoms associated with TMWD and a possible differential diagnosis for each symptom. Diagnostic audiometric findings associated with TMWD are further explored in Chap. 11.

Table 5.2 Audiologic mimickers

Signs/symptoms	Differential diagnosis
Autophony	CSOM, ETD; TMWD
Aural fullness	LFSNHL, endolymphatic hydrops/Ménière's disease, ETD, temporomandibular myofascial disorders
Pulsatile tinnitus	Vascular lesions (AVM/AVF), IIH, glomus tumors, venous hum, sigmoid sinus diverticulum, carotid pseudoaneurysm; TMWD
Conductive hearing loss	Any disorder affecting the EAC, TM or ossicular chain including otosclerosis, ETD, etc.

Imaging

The next step in the workup of a patient presenting with audiovestibular symptoms concerning for a TMWD is to obtain a high-resolution temporal bone CT scan, specifically with direct axial images, accompanied by Poschl and Stenvers reconstructions. This remains the gold standard in identifying the location of dehiscence. MRI is typically normal in SSCD but may be useful when evaluating for concurrent CSF leak or meningoencephalocele and in ruling out retrocochlear pathology [12]. Further discussion of the imaging of TMWD can be found in Chap. 12.

Differential Diagnosis for the Otologic Mimicker

Otologic Mimickers

Benign Paroxysmal Positional Vertigo (BPPV)

BPPV is the most common vestibular disorder with an incidence of 10–64 per 100,000, with an increasing 38% incidence with each decade of life [13, 14]. The disorder may be even more common than once suspected, with one study showing positive results in 9% of randomly selected geriatric patients undergoing positional testing with no former diagnosis of BPPV [15]. Vertigo provoked by position changes with quick resolution with eye fixation is the hallmark description of BPPV. 94% of cases involve the posterior semicircular canal [16]. Vertigo is typically triggered with movement towards the affected ear. A diagnostic Dix-Hallpike maneuver will incite an ipsi-directional torsional nystagmus after a short latency, resolves in 10–30 s, and diminishes with repeated positional testing (i.e., fatigues) [17]. Canalith repositioning maneuvers (i.e., Epley maneuver in posterior SSC involvement) are highly effective in treating the current episode, however does not prevent recurrent episodes, which occurs at a rate of approximately 15% per year [18]. The other semicircular canals can certainly be affected by dislodged otoconia as well. Horizontal canal BPPV is seen in up to 10% patients with positional vertigo and can be provoked with a head roll maneuver. Superior canal BPPV is considered quite rare, affecting less than 2% of positional vertigo patients. It is important to note that patients can have multiple affected canals simultaneously and if the posterior canal CRM does not resolve the symptoms, the patient should be tested for horizontal or superior canal BPPV in addition to expanding the differential diagnosis of the vertigo symptom.

BPPV and TMWD share the symptom of vertigo. The vertigo for both disorders may be provoked by head movement and in both disorders there can be a sense of disequilibrium between the vertigo attacks. However, BPPV is by definition positional and has characteristic head movements that provoke the vertigo—rolling over in bed, looking up (top shelf vertigo), and bringing the head up from a dependent position. TMWD patients do not often have a specific positioning maneuver that will induce vertigo but may describe dizziness with rapid head turning. In posterior canal BPPV, the vertigo and characteristic nystagmus will be induced by the

Dix-Hallpike maneuver. In TMWD the positioning maneuvers do not produce nystagmus but the patient may report a sense of "dizziness" when they move from supine to the seated upright position but will not have an "unwinding nystagmus" as seen with pcBPPV. Additionally, TMWD are often accompanied by auditory symptoms which are not seen in BPPV. A diagnosis of BPPV is a purely clinical diagnosis based on exam findings whereas additional diagnostic testing is required to identify the TMWD diagnosis. Failure to provoke nystagmus with positioning testing should prompt the practitioner to expand their differential diagnosis. However, one should keep in mind that the presence of BPPV or TMWD does not preclude the concomitant existence of the other.

Eustachian Tube Dysfunction

Eustachian tube dysfunction (ETD) is very common and accounts for more than 2 million patient visits per year in the adult population in the United States alone [19]; in pediatric patients, ETD is strongly associated with chronic otitis media with effusion [20]. Disorders of the Eustachian tube (ETD) can be classified as either obstructive or patulous, and both have significant symptomatic overlap with each other and TMWDs. ETD often manifests as reports of aural fullness and pressure, autophony, and muffled hearing.

Patients with obstructive ETD experience symptoms due to failure of equalization of the middle ear pressure to barometric pressure. The dynamic opening of the ET orifice in the nasopharynx allows air to travel from the nose to the middle ear; mucosal edema within the ET or anatomic variants of the ET, or both, result in failure to replenish the middle ear aeration. Failure to ventilate the middle ear cleft results in retraction of the tympanic membrane due to absorption of the nitrogen by the mucosa of the ear. Acute negative pressure on the TM is painful, as reported by those who experience barochallenged ETD (pain with air travel or scuba diving). Patients may report the need for frequent Valsalva maneuvers to forcefully open the ET in the nasopharynx and push air into the middle ear cleft.

Treatment of obstructive ETD revolves around mitigation of mucosal edema with topical steroids, ventilation tube placement, or mechanically crushing the tissue with balloon dilation. Anatomic variability can contribute to obstructive ETD but is not amenable to treatment medically or surgically. Obstructive ETD can mimic TMWDs with regard to aural fullness and muffled hearing. However, obstructive ETD patients do not experience episodic vertigo and have stigmata of chronic tympanic membrane retraction on otoscopy. Audiometrically, both groups may have a conductive hearing loss with preserved cochlear function; however, obstructive ETD patients will have abnormal tympanograms with pressures in negative excess of –150 dPa (Type C) [21].

Patulous ETD refers to an ET which is "too open." Although the atmospheric pressure is equal between the middle ear and nasopharynx, patients with patulous ETD report muffled hearing, fullness and autophony. The autophony in patulous ETD is often particularly prominent with breathing, and patients report hearing and feeling their breath in their ear. This symptom is particularly bothersome to many

patients and can sometimes be mitigated by lying flat and using gravity to pull the ET orifice closed; consequently it is important to observe the TM for respiratory movement in both the seated and the supine positions. The aural symptoms of patulous ETD can be improved by forceful sniffing to apply negative pressure to the ET orifice for temporary closure. However, the symptoms recur quickly in many patients and they may develop a habit of sniffing. Patulous ETD can occur in patients who experience a significant and rapid weight loss, such as after bariatric surgery [22]. Treatment for patulous ETD is focused on increasing tissue mass at the ET orifice in the nasopharynx and may involve irritative solutions applied via the nose (premarin nose drops) or injection of material into the torus tubarius to increase tissue bulk.

As with obstructive ETD, the symptom overlap with TMWD is considerable as both patient groups will experience aural fullness, autophony, and muffled hearing. However unlike TMWD patients, patulous patients do not report episodic vertigo, nor sound or pressure-induced symptoms. On exam, patulous patients have a normal appearing tympanic membrane on cursory evaluation; however, on closer inspection the tympanic membrane can be seen to move with the respiratory cycle. A small paper patch can be placed on the TM in the office in the patient suspected of having patulous ETD which often improves or resolves the symptoms; the added weight on the TM will not resolve the symptoms for patients with TMWD.

Ménière's Disease

Ménière's Disease (MD) is a commonly recognized cause of peripheral vertigo. Incidence ranges from 7 to 515 per 100,000 based on several studies depending on the country [23]. The classic episodic symptoms include aural fullness, fluctuating low to mid-frequency sensorineural hearing loss (SNHL), roaring tinnitus, and rotatory vertigo. Only one third of cases present with this full quadrad of symptoms, however [24]. Ménière's disease may be due to overproduction or inadequate absorption of the endolymph within the membranous labyrinth, although a full understanding of the pathophysiology is not known. In an acute attack, vertigo lasts several minutes to several hours followed by a post-vertiginous disequilibrium. Patients generally report normal balance function between episodes.

Diagnostic criteria has been set forth by the American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS) Foundation which distinguishes between definite and probable MD [25, 26]. MD is characterized by two or more spontaneous attacks of vertigo, each lasting 20 min to 12–24 h and fluctuating aural symptoms (hearing loss, tinnitus, or fullness) in the affected ear, and exclusion of other causes with testing. Definite MD has the additional criteria of audiometrically documented fluctuating low to mid-frequency SNHL in the affected ear before, during, or after an attack. ECOG findings can show an SP/AP ratio of >45%. ENG testing can show a decreased vestibular response to caloric stimulation in the affected ear. Abnormal VEMP testing can be seen with a reduction in amplitude of >40% [27]. While all of these vestibular tests may help confirm a diagnosis of MD, sensitivity is limited in all. Therefore, vestibular testing is not required for a diagnosis of MD. The AAO-HNS recently published updated Clinical Practice Guidelines

in 2020 which thoroughly dissects the prophylactic, medical and surgical treatment options available to patients with MD [28]. The overriding philosophy of MD treatment centers around noninvasive and nondestructive management for as long as possible due to the possibility of bilateral involvement.

Ménière's Disease and TMWD have significant overlap in symptomatology and exam findings. Both disorders are characterized by episodic vertigo, aural fullness, tinnitus, and hearing loss. In MD, the vertiginous episodes last for minutes to hours and the patient is often prostrate during these episodes—having severe nausea and vomiting and often unable to walk. These spells are often unprovoked and happen with little to no warning. TMWD patients usually describe a provoking trigger for their vertigo such as loud noise or straining. MD patients may notice sensitivity to salt in the diet whereas TMWD patients do not have a dietary trigger. Tinnitus associated with MD also differs in quality compared to TMWD; MD patients often describe low pitched noise in the ear (ocean, jet engine, roaring) whereas TWMD patients often have pulsatile tinnitus. The hearing loss experience by MD patients is classically a low frequency sensorineural loss with loss of clarity. By comparison, the TMWD hearing loss is low frequency but conductive in nature with preserved word understanding. TMWD can be identified on diagnostic testing (imaging and VEMP testing are most well reported) but there is no confirmatory diagnostic test for MD. There may be significant overlap between endolympatic hydrops and TMWD based on recent MRI imaging, however, the implications of these findings is not entirely clear. The reader is encouraged to further explore the association of endolymphatic hydrops, Ménière's Disease and TMWD in Chap. 18.

Otosclerosis

Otosclerosis is a disorder affecting the enchondrial bone of the otic capsule. The histopathology of this disorder is specific to the otic capsule and is termed "otospongiosis." In otosclerosis, the enchondrial bone throughout the otic capsule can undergo increased rates of bone turnover, abnormal bone deposition, and vascular proliferation [29]. Radiographically, this abnormal bone turnover results in areas of radiolucency at the fissula ante fenestram or, in patients with cochlear involvement, demineralization around the cochlear duct—referred to as a "halo sign." Otosclerosis often presents with progressive hearing loss, autophony and tinnitus. The tinnitus in otosclerosis is subjective and non-pulsatile in nature whereas TMWD patients often report pulsatile tinnitus. Patients may also have a family history of the disease or have family members who have "had surgery for their hearing." To differentiate between TMWD and otosclerosis, the practitioner may be able to utilize both physical exam findings and diagnostic testing differences. On examination, both disorders may cause tuning fork abnormalities. Also, both disorders often have a normal otoscopic examination of the tympanic membrane and middle ear cleft. In very active otosclerosis, the cochlear promontory may have increased vascularity which appears with a red hue without mass lesion (Schwartze sign). The promontory in TMWD should appear normal. On audiometric testing both groups have a conductive hearing loss. In SSCD, patients may demonstrate a supra-threshold bone line

in the low frequencies, often with closure of the air bone gap in the mid to high frequencies. Otosclerosis often demonstrate closure of their air-bone gap at 2000 Hz (Carhart's notch) due to a dip in the bone scores, whereas TMWD does not. Acoustic reflexes are particularly helpful in distinguishing the two groups. Otosclerosis demonstrates absent acoustic reflexes due to fixation of the stapes footplate whereas TMWD does not lose this reflex. Of note, there are case reports of patients having both otosclerosis and SSCD, where the SSCD was unmasked by correction of the otosclerosis [30, 31]. The reader should consider concurrent disorders if corrective surgery for otosclerosis fails to close the air bone gap or results in episodic vertigo.

Autoimmune Inner Ear Dysfunction

Autoimmune inner ear disease (AIED) occurs secondary to an immunologically mediated attack onto the audiovestibular system. Originally described by McCabe as purely audiologic in nature, patients can experiences vestibular symptoms as well [32]. AIED is uncommon, accounting for <1% of all cases of hearing loss and dizziness [32]. Symptoms include fluctuating, rapidly progressive bilateral SNHL often accompanied by tinnitus, and aural fullness. Patients are more likely to present with mild ataxia and episodic lightheadedness than true vertigo. AIED occurs more commonly in females between 20 and 50 years of age. 30% of patients with AIED will have another systemic autoimmune disorder present [33]. The inner ear is not exposed to many antigens, leading theories on the pathogenesis of AIED include cross reaction, bystander damage, intolerance, and genetic factors related to the immunologic response. Presentation can be similar to MD and TMWD but with bilateral involvement being a key distinguishing feature of AIED, though AIED can be asymmetric early in the disease process. 16% of bilateral and 6% of unilateral MD may be caused by immune dysfunction [33]. Serological testing can give mixed results [34]. Initial treatment includes systemic vs. intratympanic steroids while immunomodulatory agents are often prescribed for long-term treatment [35]. The rapidly progressive symptoms and response to steroid medications help to distinguish AIED from TWMDs.

Labyrinthitis

Labyrinthitis describes inflammation of the inner ear, commonly divided into serous vs. suppurative labyrinthitis. Suppurative labyrinthitis describes a pyogenic infection of the inner ear, which can produce severe symptoms resulting in permanent hearing loss and vestibular dysfunction and can be rapidly progressive and life-threatening if intracranial complications occur. Serous labyrinthitis describes inflammation of the inner ear without frank bacterial infection, which also can present with severe symptoms although often long-term sequela are not always observed. Both serous and suppurative labyrinthitis can be isolated to the ear (tympanogenic) or extend intracranially.

Labyrinthitis is a clinical diagnosis and can occur in the setting of acute or chronic otitis media. Inflammation can spread via an acquired pathway between the middle and inner ear spaces, notably temporal bone fracture, iatrogenic from otologic surgery, or cholesteatoma erosion (most commonly fistula formation of the horizontal SSC). However, there is often no apparent inner-middle ear communication in many cases of tympanogenic labyrinthitis. Meningitis can be a source of inflammatory and infectious spread from the meninges into the inner ear in cases of meningogenic labyrinthitis. 5–35% of patients who survive bacterial meningitis will have bilateral SNHL [36]. MRI is the preferred modality to assess patients with labyrinthitis commonly showing hyperintense labyrinthine signal on T1 post-contrast imaging sequences.

The differentiation between TWMD and labyrinthitis is based on history and audiometry. In most cases of labyrinthitis the vertigo is sudden in onset, intense in nature and continuous initially but gradually improves as central compensation occurs. The hearing loss is predominantly sensorineural in nature after resolution of a middle ear effusion; these symptoms are in contradiction to TMWD patients who experience provokable episodic vertigo and generally have a conductive hearing loss. Additionally, TMWD patients do not often experience resolution of the vestibular symptoms with time.

Mass Lesions Involving the Labyrinth

Vestibular schwannomas (VS) are tumors arising from Schwann cells within the internal auditory canal (IAC) and can present with a constellation of vertigo, SNHL, and/or facial nerve palsy depending on its location and size within the IAC. Schwann cells are also found more distally within the inner ear labyrinth itself, which can produce tumors termed intralabyrinthine schwannomas (ILS). ILS are much more rare compared to VS and are often mistaken for inflammation of the labyrinth on MRI [37]. Symptoms vary based on specific location of the ILS anatomically described by the revised Kennedy classification system [38]. Nearly all patients present with some form of hearing loss and may describe disequilibrium vs. vertigo. ILS interruption of intralabyrinthine fluid mechanics make these lesions difficult to clinically differentiate from MD and TMWD. The characteristic MRI findings of intralabyrinthine hyperintensity on post-contrast T1 weighted imaging is diagnostic [39]. Treatment is dictated by patient symptoms, tumor size, and location.

Secondary third windows are a well known complication of other masses in the temporal bone, namely cholesteatoma and petrous apex lesions. Cholesteatomas can be particularly erosive and result in fistulization of any of the labyrinthine structures although the HSCC is the most commonly involved, as the cholesteatoma sac expands into the antrum and mastoid air cells. Like TMWD patients, cholesteatoma patients will often report aural fullness, autophony, hearing loss and occasionally episodic dizziness/vertigo. Rates of occult or symptomatic fistulas in cholesteatoma vary widely with rates as high as 15% in some early series prior to routine use of pre operative imaging; current series report rates of labyrinthine fistulas between 2 and 8% [40–42]. Dizziness/vertigo in a patient with cholesteatoma is highly suggestive of a fistula [43]. The key differentiator in this group of patients is the history or

discovery of chronic inflammatory otitis media and identification of cholesteatoma. The cholesteatomatous fistula patient will most likely have an abnormal otoscopic exam belying either active disease or the stigma of prior surgery for cholesteatoma. Additionally, unlike TMWD patients, cholesteatoma patients will likely report long standing ear problems, otorrhea, non-pulsitile tinnitus, and occasionally pain.

Of note, it is possible that a patient with cholesteatoma also has an unrelated TMWD. Imaging of the temporal bone is critical in this situation as cholesteatoma surgery will not address those symptoms caused by the TWMD. To identify two distinct diagnoses, the CT imaging would need to identify the dehiscence in an anatomically separate location from the cholesteatoma and this separation would need to be confirmed at surgery as the CT images in cholesteatoma may not detect thin layers of matrix over a secondary fistula.

Petrous apex lesions may also result in otologic symptoms mimicking TMWD [44]. Cholesterol granulomas of the petrous apex will often present with aural fullness, hearing loss and dizziness. Key features that distinguish petrous apex lesions include headache, lack of episodic symptoms, and stereotypical findings on CT and MRI imaging. Other lesions of the petrous bone, such as meningioma and endolymphatic sac tumors may also present with symptoms mimicked by TMWD including aural fullness, hearing loss and vertigo. Often these symptoms are not provokable and progressive in patients with mass lesions; the hearing loss is sensorineural in nature; and they do no report autophony. Imaging will clearly differentiate a mass lesion from a TMWD.

Neurologic Mimickers

Multisensory Balance Dysfunction

Neurologic findings can vary based on the etiology of a patient's dizziness. Cranial nerves, motor and sensory findings, cerebellar testing, coordination, and mental status are all important aspects of the physical exam that may help lead towards an underlying etiology. Particular attention should be made when evaluating extraocular movements during the cranial nerve exam as previously discussed. Both motor and sensory neuropathies can contribute to vestibular symptoms. Certain neurologic findings, including dysarthria, visual disturbances, extremity weakness, or ataxia, indicate a central etiology to vertigo symptoms. Romberg and gait testing helps to assess the visual, vestibular and somatosensory coordination necessary to maintain balance.

With age and cognitive decline, the prevalence of "dizziness" increases. Dizziness is the most common complaint among patients older than 75 years presenting to a doctor's office [45]. Many studies have revealed age-related changes in the vestibular organs, together coupled with peripheral neuropathy, decreased visual acuity, impaired cognitive function, and a decline in neuroplasticity. All of these changes contribute to the increased prevalence of this multisensory balance dysfunction—"dizziness." Increased fall risk and the associated morbidity and mortality that comes with falls are a serious health concern in the elderly population. However, the misconception that all dizziness in the elderly population is age-related can result in a

delay in diagnosis of treatable etiologies [46, 47]. A thorough evaluation for treatable causes of dizziness in the aging population is imperative, along with appropriate referrals for non-peripheral causes. Many patients will benefit from practical interventions such as vestibular physical therapy, exercise programs, and falls risk reduction.

Migraine

Vertigo and migraine are two common neurologic complaints often coexistent in the general population. In one large population-based study, the lifetime prevalence of migraine was 14% and vestibular vertigo 7%, giving an expected absolute chance coincidence of 1%, though actual coincidence was found to be 3.2% [48]. Significantly less patients with tension headache reported vertigo compared to patients with migraine, 8% vs. 27% respectively [49, 50]. Many different subtypes of migraine exist including generalized with or without aura, ocular, menstrual, abdominal, vestibular, and migraine without headache. Many patients with TMWD also have coincident migraines, but this may be simply related to the high prevalence of migraines in the general population. For some patients, TMWD symptoms can be migraine triggers. Dietary and environmental triggers can be present in all types of migraine.

Vestibular migraine is the second most common cause of vertigo and the most common cause of spontaneous episodic vertigo. The description of a patient's vertigo can be spontaneous and positional along with ataxia of variable duration lasting seconds to days. Most episodes of vertigo have no sequential relationship with the headache [50]. A diagnostic criteria for vestibular migraine was created by the Migraine Classification Committee of the International Headache Society and is included below:

1. At least 5 episodes fulfilling criteria 3 and 4 (listed below).
2. A current or past history of migraine without aura or migraine with aura.
3. Vestibular symptoms of moderate or severe intensity, lasting between 5 min and 72 h.
4. At least 50% of episodes are associated with at least one of the following three migrainous features:
   (a) Headache with at least two of the following four characteristics:
      - Unilateral location
      - Pulsating quality
      - Moderate or severe intensity
      - Aggravation by routine physical activity
   (b) Photophobia and phonophobia
   (c) Visual aura
5. Not better accounted for by another ICHD-3 diagnosis or by another vestibular disorder.

Migraine is a clinical diagnosis and treatment focuses around dietary modifications, trigger avoidance and pharmacologic therapy, both prophylactic and abortive. Suspicion or diagnosis of migraine should prompt a neurology referral.

Migraine and TMWD disorders have significant symptomatic overlap but the patient history will give clues to help differentiate the two disorders. Additionally, because migraine has a high prevalence in the population, there is a strong possibility that both patient groups can present with aural fullness, tinnitus, and episodic balance dysfunction. Tinnitus in migraine disorders is non-pulsatile and may be unilateral or bilateral, whereas TMWD patients more often experience pulsatile tinnitus in just the affected ear. Often the balance dysfunction in migraine is variable in its manifestation such that patients may have both episodic true vertigo as well as a sense of disequilibrium at different times. Balance dysfunction in migraine does not have to occur temporally associated with head pain, however, other migraine associated symptoms are associated as noted above in the IHS criteria. The balance dysfunction of TMWD is not associated with headache but the associated symptoms of hyperacusis/phonophobia and nausea/vomiting are similar between the two disorders. Often patients will have symptoms consistent with both migraine and TMWD. In these cases, optimal control of migraine is imperative prior to consideration of surgical treatment of TMWD when they are coexistent. Failure to treat common migraine or vestibular migraine prior to surgery may result in prolonged recovery times or overt surgical failure to treat the TMWD associated balance dysfunction [51]. We strongly recommend maximal medical treatment of migraine in those with concomitant TWMD prior to any attempts at repair of the dehiscence. Further exploration of migraine disorders and SSCD can be found in Chap. 25.

Mal de Débarquement Syndrome

Mal de Débarquement (MDD) is characterized by the persistent feeling of dizziness and disequilibrium lasting longer than one month after prolonged sea voyage though can occur after air travel, train rides, space flight, and even skiing. MDD should be distinguished from land sickness, which is much more short-lived resolving spontaneously within two days. In the majority of patients with MDD, symptoms are not experienced until after disembarking [52]. MDD affects mostly females between 30 and 50 years of age with a mean duration of 3.5 years [53]. The pathophysiology of MDD remains controversial and in general is considered a variant of motion sickness, though this does not explain the female and age predominance. Others believe it to be related to migraine or a form of anxiety. Treatment is often medical and largely ineffective, aimed primarily at keeping the patient comfortable until spontaneous remission [52]. MDD has a clear inciting/index event and the majority of TMWD do not (traumatic TMWD being the exception). MDD symptoms are constant and can be perceived at rest and in motion. TMWD patients may have some disequilibrium, however the majority have episodic vertigo which does not improve over time and many have audiologic symptoms as well, which MDD does not.

Psychiatric Mimickers

Prior to Minor and colleagues' landmark description of SSCD in 1998, patients suffering from SSCD and other TMWDs were inappropriately labeled "crazy" and "difficult" as they did not fall within the categories of known vestibular disorders at that time [3]. Patients with TMWDs can still face these preconceived designations due to its relatively new and unknown familiarity amongst primary care providers. The DSM-V criteria for "panic attack" include four or more of the following symptoms which occur suddenly and are accompanied by fear or a "sense of discomfort" [54]:

1. Palpitations, pounding heart, or accelerated heart rate
2. Sweating
3. Trembling or shaking
4. Sensations of shortness of breath or smothering
5. Feeling of choking
6. Chest pain or discomfort
7. Nausea or abdominal distress
8. Feeling dizzy, unsteady, lightheaded, or faint
9. Derealization (feelings of unreality) or depersonalization (being detached from oneself)
10. Fear of losing control or "going crazy"
11. Fear of dying
12. Paresthesias (numbness or tingling sensation)
13. Chills or hot flushes.

Many of the symptoms above accompany TMWD as well, so when patients present to the emergency room and report dizziness, nausea, disorientation and a feeling of "going crazy," the TMWD patient will often receive the wrong diagnosis. Additionally, it is easy to imagine how descriptions like "hearing my eyeballs move" and other internal bodily sounds may trigger a psychiatry referral amongst physicians unaware of the TMWD entity. Anecdotally, many patients experience a sense of relief to the anxiety surrounding their symptoms when the correct diagnosis of a TMWD is made. A careful otologic history in these patients can often differentiate panic attacks from TWMD. TMWD patients may certainly experience nausea, palpitations and sweating immediately following a vertigo episode. The timing of the vertigo in relation to the other symptoms is an important feature. Other clues to the TMWD diagnosis include a history of pulsatile tinnitus and nonfluctuating hearing loss. TMWD symptoms are often triggered by a physical activity such as straining, applying pressure to the ear canal or exposure to loud impulse noise. Panic attacks may have no inciting event or can be triggered by intrusive thoughts. There is certainly overlap between TMWD and anxiety as many patients will avoid, or have significant anxiety about, activities which have triggered symptoms in the past. Thus TMWD can cause or exacerbate preexisting anxiety and panic disorders [51]. The reader can explore patient stories about the TMWD journey in Chap. 27. Unfortunately for these patients, incorrect psychiatric diagnoses are still commonplace.

Table 5.3 lists the comparisons of similar and different symptoms and findings for the disorder mimicked by TMWD.

Table 5.3 Symptom differentiation for mimicking disorders

Mimicker disorder	Similar symptoms	Different symptoms/signs
Ménière's Disease	Aural fullness, episodic vertigo, muffled hearing	Non-pulsatile tinnitus, sensorineural hearing loss, no autophony
Otosclerosis	Conductive hearing loss, normal tympanogram findings	Absence of vertigo, absent acoustic reflexes
Obstructive Eustachian tube dysfunction	Aural fullness, autophony, conductive hearing loss	Absence of vertigo, abnormal otoscopic exam findings, abnormal tympanogram findings
Patulous Eustachian tube dysfunction	Aural fullness, autophony	History of rapid weight loss, absence of vertigo, respiratory mobility of the TM on otoscopy
BPPV	Positional vertigo, disequilibrium	Absence of hearing loss, no aural fullness, no autophony
Labyrinthitis	Vertigo, hearing loss	Rapid onset of symptoms, unilateral SNHL
Autoimmune inner ear disease	Vertigo, hearing loss	Rapid onset of symptoms, responsive to steroids
Secondary labyrinthine fistulas	Vertigo, aural fullness, hearing loss	History of ear disease, characteristic imaging findings, abnormal otoscopic exam
Schwannomas	Vertigo/disequilibrium, hearing loss	SNHL, characteristic imaging findings, non-pulsatile tinnitus
Migraine	Episodic vertigo/disequilibrium, tinnitus	Headaches, non-pulsatile tinnitus, dietary triggers
Mal de Débarquement	Disequilibrium	History of sea voyage/inciting event, no aural fullness, no hearing loss, no autophony, no hearing loss
Multisensory balance dysfunction	Disequilibrium	Absence of hearing loss, vision changes, peripheral neuropathy
Panic disorder	Somatic symptoms with events, anxiety	Triggered by intrusive thoughts

Asymptomatic Labyrinthine Dehiscence

The true incidence of labyrinthine dehiscence is hard to quantify. It is possible that a patient may have several of the symptoms of TMWD and a dehiscence seen on radiography but whose symptoms are not caused by the radiographic dehiscence. Additionally, there are certainly patients who have a radiographically identified dehiscence who do not have symptoms of TMWD. Several studies including an examination of 1000 adult temporal bones by Carey et al. revealed a 0.5% incidence of dehiscence and an additional 1.4% incidence of markedly thinned bone overlying the SSC [55]. Similar radiographic studies have demonstrated a 3–9% rate of radiographic SSCD though this is likely overestimated due to resolution limitations and absence of Poschl or Stenvers reconstructions [56–59]. Others have reported a 3% rate of radiographic dehiscence but just 0.6% had clinical manifestations consistent

with SSCD [59]. The incidence/prevalence of symptomatic SSCDs is unclear, and the incidence of other less common TMWDs is even less clear as most descriptions of other windows are limited to case reports or small case series. It is possible that TMWD symptoms may occur on a spectrum, though further studies are necessary to determine what factors make a labyrinthine dehiscence symptomatic vs. asymptomatic.

Ockham's Razor

In training, we are often told to not make two diagnoses when one will suffice. However, it bears mentioning that TMWD can exist concurrently with any of the above mentioned disorders. For example, there is emerging evidence that many patients with SSCD also have hydrops identified on MRI. The prevalence of migraine is quite high in the general population and many symptoms of migraine overlap with TMWD. The literature is rife with reports of patients undergoing stapes surgery only to develop TMWD syndrome after the oval window fixation is corrected because they had an undiagnosed dehiscence somewhere else in the otic capsule. It is important to try to identify which disorder is causing the primary symptom for the patient and attempt treatment for that disorder. For instance, vestibular migraine should be controlled before surgery for a radiographic SSCD, as untreated vestibular migraine will likely lead to symptomatic failure of SSCD surgery.

Bilateral Third Mobile Windows

The congenital theory of SSCD argues thin bone overlying the SSC either causes a persistent dehiscence vs. predisposition to dehiscence later in life. This may explain why up to 50% of patients with SSCD will have bilateral defects [59]. Those with bilateral SSCD may develop oscillopsia [8]. Many patients are only symptomatic on one side, therefore treatment should center around addressing the more symptomatic ear and monitoring for resolution of symptoms before possibly proceeding with treatment of the contralateral.

Diagnostic Algorithm

When a patient presents for evaluation of various otologic complaints, it is helpful to organize the workup oftentimes on the most prominent symptoms experienced by the patient. Figure 5.1 outlines possible workup algorithms based on the most bothersome/prominent symptom reported by the patient on presentation.

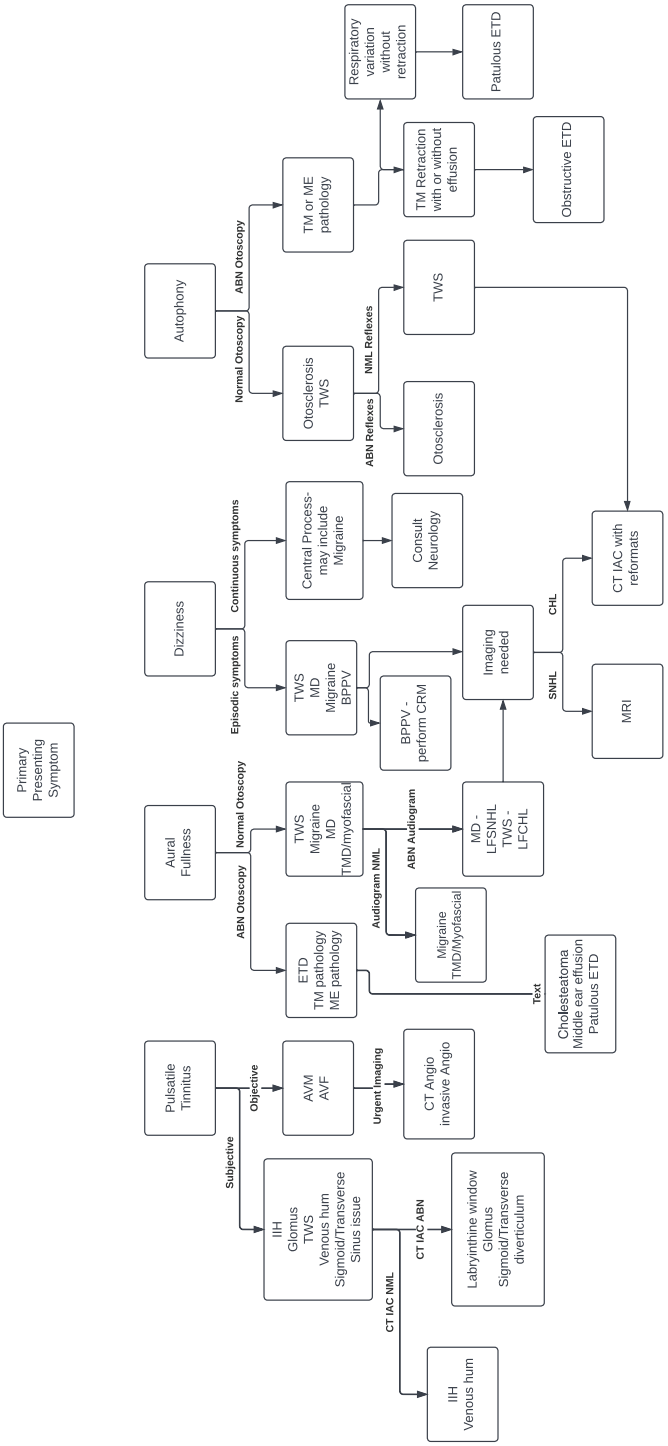

Fig. 5.1 Workup algorithms

Conclusion

Third window syndrome has a variety of presenting symptoms and signs. There is no "one thing" that points the practitioner to the correct diagnosis but a constellation of symptoms and findings that, when taken together, suggest the correct course of action. Practitioners must maintain a high index of suspicion for TMWD in patients who present with audiovestibular complaints; the differential diagnosis is broad but with a careful evaluation the correct diagnosis can be made.

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