Diagnostic Issues in Tinnitus: a Neuro-otological Perspective

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ABSTRACT

Arriving at a diagnosis to account for tinnitus begins with the patient's description of the percept, because in some cases the quality of the tinnitus will make the diagnosis (e.g. clicking) and in other cases it will give direction to the diagnostic evaluation (e.g. pulsatile). With the exception of dural arteriovenous malformations, the source of pulsatile tinnitus can be determined without conventional cerebral angiography.

Establishing a diagnosis for non-specific tinnitus is difficult because (i) tinnitus is common in the general population, (ii) for any disease not all subjects will develop tinnitus, and (iii) tinnitus can be multi-factorial. Unilateral tinnitus with non-specific characteristics must be suspect for an acoustic neuroma. In the absence of stress or intense sound exposure, if tinnitus (a) is intermittent, (b) fluctuates widely in loudness or location, or (c) has a diurnal pattern, then somatic influences upon tinnitus from the head or upper cervical region should be suspected .

Key words: Somatic (craniocervical) tinnitus, dorsal cochlear nucleus; hallucinations

Tinnitus is the perception of sound in the absence of an external sound. As such it is a symptom and may have many diverse causes. The purpose of this chapter is to describe an approach to establishing the origin of this symptom. For this reason, we use the presenting complaint as the organizing principle, since this is what confronts the clinician upon encountering the tinnitus patient.

The evaluation of a tinnitus complaint should include the standard elements of any medical evaluation with close attention to the circumstances surrounding the onset of the tinnitus including any association with new medications, psychosocial stressors, a concurrent medical illness, other auditory, vestibular or neurological complaints or a head, neck or dental disorder. Exacerbating and remitting factors should be sought including diurnal variations in the tinnitus. The most important features of the tinnitus percept that must be ascertained are its (1) quality, particularly whether or not it is pulsatile, (2) its location, whether it is heard in one ear or not, (3) its variability, whether it is intermittent or constant, and (4) its pitch, whether it is predominantly low or high frequency in character. In addition to the standard otological physical examination, in general the tinnitus exam should include inspection of the teeth for evidence of bruxism, listening around the ear and neck for sounds similar to their tinnitus, and palpation of the cranio-cervical musculature for muscle tension and tenderness with special attention to any asymmetries. All subjects should have a recent audiogram.

TINNITUS QUALITY: SPECIFIC

Some types of tinnitus have such characteristic features that the description alone is the major determinant of the direction of the diagnostic approach. These types of tinnitus can be described as strictly in one ear or non-lateralized. Patients use a variety expressions to describe non-lateralized tinnitus, such as: "both ears about equally," "both ears but worse in one ear," "in the head but not strongly toward either side," or "in the head toward one side." Aside from hallucinations all tinnitus with a characteristic quality are "somatosounds" - physical sounds generated by the body and heard by one or both ears.

Tinnitus Always Unilateral

Coarse intermittent sounds coincident with jaw or head movements: Such tinnitus is typical of a foreign body such as cerumen, water (or other liquids), or a hair resting against the tympanic membrane. Inspection of the ear canal will reveal the source.

Fluttering . Stapedius muscle contractions tend to be described as a fluttering. If the fluttering is associated with facial movements, then stapedial contractions is highly likely to be causing the fluttering sound. This is most commonly seen chronically after recovery from Bell's palsy, unilateral facial paralysis. When the affected side of the face contracts, the ipsilateral stapedius muscle also contracts (synkinesis) due to aberrant facial nerve regeneration. Abnormalities in the pattern of the stapedial reflex or acoustic impedance measurements corresponding to the characteristics of the patient's tinnitus can occur (Marchiando, Per-Lee et al. 1983) . When fluttering unilateral tinnitus occurs in isolation with no other associated signs or symptoms, no further diagnostic studies are required.

Tinnitus Maybe Unilateral or Non-lateralized

Pulsatile

Whether or not the tinnitus is pulsatile should be established at the outset of the evaluation. If described as pulsatile, then the next step is to determine whether it is related to the cardiac cycle. This can be evaluated by comparing the examiner's silent count of the patient's cardiac pulse while at the same time the patient is silently counting the pulsations of his tinnitus. The examiner indicates when the counting interval starts and stops and then the two counts are compared. If the counts are virtually identical, then the pulsatile tinnitus is cardiac related and a vascular source must be sought. However, if the two counts are discordant then the tinnitus is not cardiac related and other causes must be considered.

The patient's history can give clues to the source of the pulsatile tinnitus (Sismanis and Smoker 1994) . An association with headaches, blurring of vision, and menstrual irregularities in an obese woman is suspicious for benign intracranial hypertension. Abrupt onset with unilateral neck or head pains suggests a carotid dissection. Changes in tinnitus intensity with head turning suggests a venous source for the tinnitus --- from a source ipsilateral to the direction that decreases the tinnitus. If the patient can obliterate the tinnitus with localized pressure in the periauricular region then an emissary vein is probably accounting for the tinnitus. An associated fluctuating hearing loss raises the possibility of microvascular compression of the auditory nerve causing the pulsatile tinnitus (Ohashi, Yasumura et al. 1992; Waldvogel, Mattle et al. 1998) . However, whether or not vascular compression of the auditory nerve can cause tinnitus has never been convincingly established; MRI studies of patients with unilateral tinnitus can detect vascular compression of the auditory nerve on the asymptomatic side as frequently as on the symptomatic side (Makins, Nikolopoulos et al. 1998) .

The physical exam can provide key information about the pulsatile tinnitus, as well. A crescentic purple coloration to the tympanic membrane is diagnostic of a glomus jugulare tumor. Otoscopic observation of a red mass behind the tympanic membrane is evidence for an aberrant carotid artery, dehiscent jugular bulb, or a vascular tumor. A unilateral conductive hearing loss in association with ipsilateral pulsatile tinnitus and an otherwise normal exam suggests otosclerosis, as does Schwartze's sign (a red hue behind the tympanic membrane on otoscopy). Detection of a bruit ipsilateral to the pulsatile tinnitus suggests that the tinnitus is from the same source as the bruit. The source of the bruit then must be sought. If localized to the region of the carotid artery bifurcation, then fibromuscular dysplasia, carotid stenosis or carotid dissection are suspected; an associated ipsilateral Horner's syndrome would suggest a carotid dissection. If more widely distributed such as throughout the periauricular region or even more widespread a dural arteriovenous fistula becomes likely. If heard over the globe a carotid-cavernous sinus fistula is suspected, particularly if there should be an associated proptosis. Obliteration or reduction in the intensity of the pulsatile tinnitus with ipsilateral jugular compression (light or moderate pressure below the angle of the jaw) implicates a venous source of the tinnitus; whereas a decrease in the tinnitus with ipsilateral carotid compression implicates an arterial source arising from the carotid system. If venous pulsations are seen within at least one of the optic fundi, then cerebrospinal fluid pressure is normal and raised intracranial pressure can be ruled out.

The diagnostic studies following the initial visit will be guided by the findings of the clinical evaluation and laboratory studies (figure 1). Because high cardiac output states such as anemia or hyperthyroidism can cause pulsatile tinnitus (usually bilateral), all patients should have a thyroid profile and a hematocrit. If a carotid lesion is suspected then either a duplex ultrasound study of the carotid or MRA should be performed. If a retrotympanic mass is suspected, then a high-resolution contrast-enhanced CT scan of the temporal bones should be obtained. Otherwise a contrast-enhanced MRI scan of the temporal bone and cranium should be obtained. The MRI scan may not detect anomalous arterial patterns such as a persistent stapedial artery, so a non-contrast high resolution CT scan of the temporal bone is performed should the MRI scan be normal. If still no etiology is apparent and neither papilledema nor retinal venous pulsations were observed [by the examiner and/or a neuroophthalmological consultant] then cerebrospinal fluid pressure should be measured via a lumbar puncture. If all the above non-invasive imaging studies have been unremarkable and raised intracranial pressure has been ruled out, then cerebral angiography should be considered, because a dural arteriovenous malformation can sometimes go undetected by any other diagnostic study, even though there may or may not be a thrill or bruit on physical examination (Waldvogel, Mattle et al. 1998; (Weissman and Hirsch 2000) . Because significant, but rare, morbidity can occur with angiography, careful deliberation must be given to the decision to proceed with angiography.

Figure 1.

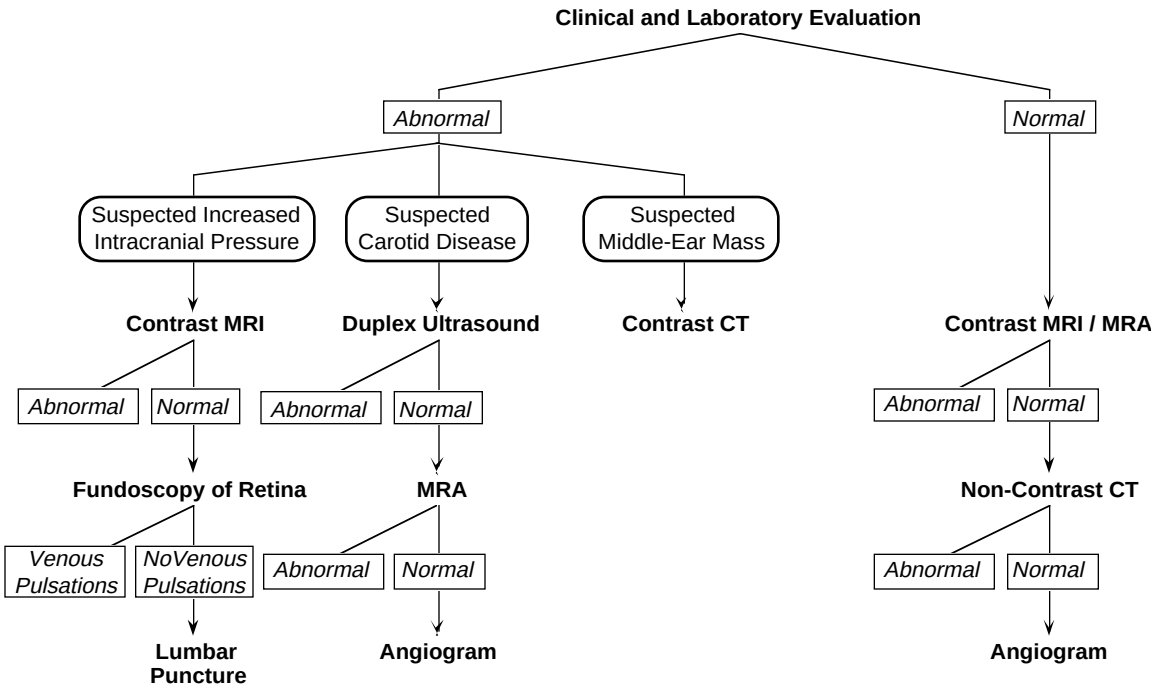

Figure 1. Pulsatile tinnitus: diagnostic algorithm. Laboratory studies should include a hematocrit and thyroid profile. MRI = magnetic resonance imaging. MRA = magnetic resonance angiography of cervical and intracranial vasculature. CT = thin section computed tomography of temporal bone. After Sismanis and Smoker (1994).

Clicking A relatively rare form of tinnitus, clicking tinnitus appears to be due to contractions of tensor tympani or the nasopharyngeal muscles controlling the patency of the Eustachian tube [dilator tubae, salpingopharyngeus, tensor veli palatini]. It can sometimes be bilateral as well, in which case, it is usually associated with palatal myoclonus. The diagnosis can be established by inspection of the nasopharynx either directly or with nasopharyngoscopy for muscle contractions coincident with the clicking. Often the clicking can be heard by the examiner listening carefully at the external auditory canal either with his ear, his stethoscope, or a low-noise microphone system placed in the external auditory canal such as is used for measuring otoacoustic emissions. Sometimes acoustic impedance measurements have abnormalities that correspond to this type of tinnitus or inspection of the tympanic membrane can reveal movements coincident with the clicking.

Autophony (echoing of the voice), or blowing tinnitus. The characteristics of this type of tinnitus are so unique that the history alone virtually makes the diagnosis, namely a patulous ipsilateral Eustachian tube. Patients describe blowing sound with respiration and an echoing quality to their own voice. Confirmatory features include disappearance of their complaints when their head is in a dependent position and abnormally large changes in the tympanic membrane acoustic impedance with respirations.

Hallucinations (non-verbal, stereotyped repetitive). Unlike the hallucinations associated with psychoses, these patients have no associated thought disorder, and they are not hearing voices with meaningful, personally relevant content. Rather the hallucinations are either "musical" in which patients report hearing one or a series of familiar tunes incessantly, or "auditory" in which a variety of different sounds are described (Obach and Obach 1996) .

Typically the strictly musical hallucinations occur in elderly patients (more commonly in women) with a longstanding progressive moderate to severe bilateral hearing loss. The tunes can be vocal and/or instrumental. While they are usually bilateral, they can be unilateral even with a bilateral hearing loss. Occasionally they can be precipitated by a new medication, and the hallucinations resolve when the medication is stopped. If the presentation is typical, no brain imaging is necessary.

Auditory hallucinations differ from musical hallucinations in several respects. They are usually abrupt in onset and associated with focal neurological findings due to a brainstem stroke or space occupying lesion. There usually is no major preexisting chronic hearing loss and the hallucinations are usually not only musical, but may have a variety of other sounds such as bells or a water fall. They often are transient (Obach and Obach 1996) . Brain imaging is required.

TINNITUS QUALITY: NON-SPECIFIC

Included in this category are a variety of descriptors of tinnitus such as buzzing, tonal, hissing, humming, ringing, roaring, rushing, whistling and whooshing, crickets, etc. None of these descriptors point to a specific diagnosis. Roaring while non-specific is often associated with Ménière's syndrome. Since the quality of the tinnitus is non-specific, aids in making a diagnosis must come from sources other than how the tinnitus sounds. Associated symptoms, circumstances surrounding the onset of the tinnitus, and ameliorating and exacerbating factors are some of the pointers to the diagnostic entity that is accounting for the tinnitus symptom.

Table 1.

Non-specific tinnitus: Reasons for difficulties in establishing a diagnosis
1. Tinnitus is common in the general population
2. For any disease, not all subjects will develop tinnitus
3. The coexistence of tinnitus and a disease does not imply that the two are related
4. Tinnitus can be multi-factorial

In attempting to arrive at a diagnosis for tinnitus in which the characteristics of tinnitus are non-specific, there are several important considerations that must be kept in mind [see table 1]. The first is that tinnitus is very common in the general population. Many people have tinnitus but have never complained of tinnitus and have not sought any medical attention for it. Heller and Bergman (1963) found such tinnitus in 94% of people they studied. In our recent study in which normal subjects were placed in a low-noise room, 55% reported having ongoing tinnitus, but only 20% had been previously aware that they had any tinnitus. Besides this chronic ongoing tinnitus, 85% of these subjects reported having experienced another type of "normal" tinnitus; namely, transient (typically less than a minute) unilateral tonal tinnitus with a blocked feeling of the same ear. Another type of commonly experienced transient tinnitus follows exposure to loud sound. About 55% of this population recalled such tinnitus, lasting from a few minutes to several hours or even days. In fact, patients have occasionally presented to our clinic complaining of one of these types of "normal" tinnitus.

A second important consideration is that for any pathological process that is well known to be associated with tinnitus, not all subjects with this diagnosis will in fact develop tinnitus. In surveys of profoundly deaf subjects, about 80% will have tinnitus, but 20% have no tinnitus (Levine 1999a) . Hence, the presence of tinnitus and a pathologic process by itself does not imply that the two are related. Because there is no obligatory association between the pathologic process and tinnitus, it remains a possibility that even though a tinnitus patient has a disease known to be associated with tinnitus, his tinnitus may not be related to the disease; rather the tinnitus and the pathologic process could coexist but be unrelated.

Because tinnitus is common in the general population, a third consideration is that the pathological process only draws the patient's attention to his pre-existing tinnitus. Furthermore, if the disease in and of itself did not cause the tinnitus, but hearing loss is part of the disease, the hearing loss could be "unmasking" the pre-existing tinnitus. Just as bringing a normal subject into a low-noise environment, can make the subject aware of tinnitus they had not appreciated previously, so could a hearing loss unmask tinnitus. Therefore, the question always remains in any patient with non-specific tinnitus, whether the tinnitus was pre-existing and unmasked either due to (a) his loss of hearing, or (b) his attention being drawn to his hearing.

Since on the one hand non-specific tinnitus can be physiological, and on the other hand non-specific tinnitus is not obligatorily associated with any pathologic process, establishing a diagnosis that is accounting for any tinnitus is more problematic than for most medical symptoms.

In general, there is a level of confidence associated with any diagnosis that might account for a symptom. For tinnitus some relationships increase this confidence. The first is a temporal association between the tinnitus and the diagnostic consideration. For example, in Ménière's syndrome, if the tinnitus fluctuates with the hearing loss and vertigo, then this strengthens the confidence of the association between the tinnitus and Ménière's syndrome. Another way the confidence of the diagnosis is strengthened is if the pitch of the tinnitus corresponds to the audiometric hearing loss pattern. Considering again Ménière's syndrome, where early in the illness low-frequency hearing loss often predominates, if the tinnitus is described as roaring and/or the pitch match is a low frequency one, then the confidence of the relationship between

the Ménière's syndrome and the tinnitus is strengthened. Likewise, changes in the tinnitus percept that are closely coupled to changes in physical findings such as described in Cases 1 and 2, strengthens the association between the two.

Another consideration in attempting to establish a diagnosis to account for a patient's tinnitus is that the cause of the tinnitus may be multi-factorial. Tinnitus can be considered a threshold phenomenon, such that while any one factor, such as chronic progressive hearing loss, may not be sufficient to elicit a tinnitus complaint, two or more factors may synergistically lead to the tinnitus becoming symptomatic (see Cases 2 and 3).

Closely related to the threshold idea is the concept of "triggering factors" that can lead to symptomatic chronic tinnitus (Fowler 1943; Coles 1996). Such factors include psychosocial stress, viral infection or the post-viral state, medications or withdrawal from medications, and head and neck somatic factors (whiplash, temporomandibular joint syndrome, ear syringing, trauma not involving the head, etc.). Clinical experience suggests that the clinical problem of tinnitus can be precipitated by one (or more) of these triggering factors (Case 4). While a triggering factor may appear to be responsible for initiating the tinnitus, sometimes the tinnitus will persist despite resolution of the triggering factor.

If, as often occurs, none of these confidence-raising factors are evident, any association with a preexisting condition such as chronic hearing loss should be considered very tenuous. In such a case, the most conservative diagnosis is "idiopathic, possibly related to condition X". As Coles writes, "If the probability is assessed as being over 50% that a particular condition is causing the tinnitus, ... most cases of tinnitus would have to be classified as 'unknown'" (i.e. idiopathic) (Coles 1996) .

With these considerations in mind, we next consider diagnostic entities that appear to be associated with non-specific tinnitus.

Tinnitus Always Unilateral

Never with vestibular symptoms

Conductive hearing loss. Any type of unilateral conductive hearing loss, such as cerumen impaction, ossicular discontinuity, or otosclerosis, can be associated with tinnitus of that ear. The tinnitus may be related at least in part to an unmasking of a "normal" underlying tinnitus, as discussed above. Otosclerosis may sometimes be associated with inner ear involvement which could be contributing to the tinnitus as well.

Otoacoustic emissions. While spontaneous otoacoustic emissions are common (75% of female and 45% of male normal or near normal ears), tinnitus due to spontaneous otoacoustic emissions is uncommon; it is said to be accounting for the tinnitus of 1-2% of the patients of one British tinnitus clinic. The diagnosis is made by measuring an emission and showing that its suppression abolishes the tinnitus. The emission can be suppressed in either of two ways: (a) presentation of a low-level tone near the emission frequency or (b) the use of aspirin (Penner and Coles 1992) . Some anecdotal reports suggest that trigger factors such as noise exposure might be associated with some cases of tinnitus thought to be related to otoacoustic emissions (Coles 1996) .

May be with vestibular symptoms

Ménière's Syndrome. As a syndrome this condition is in fact defined by tinnitus as one of its cardinal features. The full-blown picture consists of episodic attacks of intense vertigo persisting hours, fluctuating unilateral hearing loss typically involving the lower frequencies (in the early stages), ear fullness and a roaring low-frequency tinnitus. Early in its course there may be no persistent symptomatology; however, with recurrent episodes any or all of the symptoms can persist and cumulatively progress with each recurrence. While the tinnitus is often described as roaring early in the illness, with more advanced stages of the syndrome, the tinnitus tends to become more variable in its description. There are no definitive tests to establish the diagnosis; however, electrocochleography can be supportive of the diagnosis, if there is a large ratio of summating potential amplitude to action potential amplitude. Documentation of the fluctuation in hearing with serial audiograms is likewise supportive of the diagnosis. The FTA-abs test can sometimes be positive in this syndrome.

Formes frustes of this syndrome may occur. In particular episodic low frequency fluctuating hearing loss with a contemporaneous roaring tinnitus and aural fullness may occur without vertigo.

Perilymphatic fistula. Like Ménière's syndrome hearing loss, vertigo, and tinnitus may occur together. However, the tinnitus and hearing loss tend to be high frequency (hissing, crickets, etc.) with no recovery. The fistula consists of a communication between the perilymph of the inner ear fluids and the middle ear through a round or oval window defect or sometimes a defect of the bony labyrinth. The defect can be caused by barotrauma (e.g. airplane descent or ascent from diving), head trauma, valsalva, or erosion of the bony labyrinth due to an inflammatory or neoplastic process, or following middle ear surgery such as stapedectomy. The diagnosis can be suggested by the “fistula test" - the induction of nystagmus by positive

or negative pressure applied to the external auditory canal. If symptoms persist, and the findings are suggestive then the middle ear can be explored for a fistula with patching of the round and oval windows. Generally the hearing loss and tinnitus are not improved by patching the oval and round windows whether or not a leak is found at surgery.

Herpes Zoster Oticus (Ramsay-Hunt Syndrome) Intense ear pain followed by ipsilateral tinnitus, hearing loss, vertigo and facial paralysis will be recognized as due to herpes zoster once vesicles appear on the pinna, external auditory canal, or tympanic membrane.

Cerebellopontine angle tumors. The most common presentation of such tumors is a gradual unilateral sensorineural hearing loss with minimal if any vestibular complaints. Dizziness and facial weakness, in general, are either non-existent or very minor accompaniments of acoustic neuromas at any time. The audiometric pattern is variable. They are more likely to have poor speech discrimination, acoustic reflex decay, and pure tone decay. Other presentations do occur including (a) unilateral tinnitus only or (b) sudden hearing loss with or without subsequent recovery.

Any unilateral tinnitus with or without unilateral sensorineural hearing loss must be considered suspect for a cerebellopontine angle tumor, even though only about 3% of all patients with acoustic neuromas first present complaining of unilateral tinnitus only (Dornhoffer, Helms et al. 1994; (Morrison and Sterkers 1996) . Therefore, should every such patient have a contrast-enhanced MRI scan to exclude the diagnosis? In 14 years of evaluating all patients with unilateral tinnitus for a cerebellopontine angle lesion, we have never detected a lesion of the cerebellopontine angle. Dawes and Basiouny have reported a similar experience; one acoustic neuroma was detected in 174 patients with unilateral tinnitus (Dawes and Basiouny 1999) . Alternatives to obtaining the "gold standard" contrast-enhanced MRI scan immediately are (1) using the short-latency brainstem auditory evoked potentials, a good but imperfect test, to decide whether or not to proceed to the MRI scan, or (2) following the patient with revisits and repeat audiograms at 6 month intervals, looking for development of an ipsilateral hearing loss before going to the MRI scan. The issue ultimately becomes one of cost-effectiveness and availability of MRI scanning, since there is virtually no morbidity associated with the MRI scan. As such, it will depend upon local factors. A definitive study to answer this question for any locality has not been done (Dawes and Basiouny 1999) ; however, one study suggests that an MRI protocol tailored to rule out a cerebellopontine angle lesion could be highly cost-effective (Carrier and Arriaga 1997) .

Central nervous system - caudal to trapezoid body The hallmark of tinnitus due to a disorder of the central nervous system is other neurological system involvement. If the central auditory nervous system involvement occurs between the ear and the trapezoid body (where the auditory inputs from the two sides intermix and cross) then there may be an associated ipsilateral hearing loss with unilateral tinnitus. The type of associated neurological involvement will depend crucially upon the location of the lesion. With unilateral tinnitus it can include dizziness, diploplia, limb ataxia, ipsilateral facial weakness, ipsilateral facial paresthesias, but contralateral limb paresthesias. The tinnitus is often transient. Intrinsic or extrinsic neoplasms, stroke, demyelinating disease, inflammatory diseases, meningitides can all lead to unilateral tinnitus and hearing loss. The diagnosis will be established by the pattern of neurological system involvement, the temporal profile of the illness, and the results of ancillary diagnostic studies such as MRI scanning, cerebrospinal fluid examination, or arteriography.

Sudden idiopathic hearing loss. Patients can present with an abrupt unilateral tinnitus as their only complaint and upon evaluation are found to have a corresponding unilateral hearing loss of which they may have been unaware. More typically they complain of both the hearing loss and tinnitus or hearing loss alone. The hearing loss and tinnitus are abrupt in onset and always unilateral. The quality of the tinnitus usually is closely related to the pattern of the pure tone audiogram. Vestibular symptoms, should they occur, are usually not prominent. Once other rare but identifiable causes have been considered, such as cerebellopontine angle tumor, cochlear ischemia, syphilis, or herpes zoster, then the diagnosis of sudden idiopathic hearing loss is secure.

Tinnitus Maybe Unilateral or Non-lateralized

Always with hearing loss

Acute acoustic trauma. There is no difficulty establishing the diagnosis of tinnitus due to acoustic trauma when the history is one of immediate development of hearing loss and tinnitus following an intense sound exposure with partial or complete recovery of hearing (temporary threshold shift) over a few days. On the other hand, if the audiogram is normal and the tinnitus does not immediately follow a unilateral intense sound exposure, then other causes for the tinnitus must be sought.

Chronic progressive hearing loss (presbycusis, chronic acoustic trauma, hereditary hearing loss). These three conditions can be considered together, since they affect only hearing, generally have a symmetric hearing loss, and are slowly progressive, albeit at different rates. The establishment of a causal relationship between chronic progressive hearing loss and tinnitus is problematic, because there is no perceptible change in the hearing or audiogram associated with the onset of the

tinnitus. What has been well established is that the prevalence and reported loudness of tinnitus increase with increasing hearing loss (Chung, Gannon et al. 1984) . However, for any patient with chronic progressive hearing loss and recent onset of tinnitus a triggering factor or other cause for the tinnitus must be sought (see Case 2 and 3). The association between tinnitus and chronic progressive hearing loss must be considered tenuous.

Autoimmune inner ear disease. This condition is like chronic progressive hearing loss except the progression of the hearing loss is measured in weeks or months rather than years. Due to its more rapid time course, the association of tinnitus with the disease is more compelling. Blood tests detecting inner ear antibodies support the diagnosis.

Maybe no hearing loss

Somatic (Head or Upper Cervical): Observations abound supporting the notion that head and neck somatic events can be associated with tinnitus. Prior to their first visit, about 20% of patients in our clinic have noticed that they can modulate their tinnitus somatically, such as by clenching the teeth or pushing on various places on the head. For the past two years we have been systematically examining our patients with a battery of isometric head and neck contractions (Case 1). More than 75% of patients can modulate their tinnitus. A variety of changes can occur. Most commonly the tinnitus can get louder, but many times the tinnitus can become quieter, as well, particularly if the tinnitus is unilateral (Levine 1999b) . Less frequently patients describe pitch or location changes.

Tinnitus is generally included amongst the features associated with pain in the temporal or preauricular region that goes by various names such as Costen's syndrome, craniomandibular disorder, and temporomandibular joint syndrome. Well designed studies have shown a higher incidence of tinnitus in normal hearing subjects with temporomandibular joint syndrome than in controls (Chole and Parker 1992) . The same is true regarding whiplash (Tjell, Tenenbaum et al. 1999) . From multiple other observations and case reports, the concept of tinnitus associated with whiplash and temporomandibular joint syndrome can be generalized to include tinnitus associated with any disorder of the upper cervical region and head, including dental pain.

Our observations and those of others have indicated that the tinnitus temporally associated with unilateral somatic disorders are localized to the ipsilateral ear (Cases 1 and 2) (Levine 1999a) . Hence unilateral tinnitus with no associated auditory or vestibular symptoms such as hearing loss, must be suspect for an ipsilateral head or neck somatic disorder. The physical examination should include (1) inspection of the teeth for evidence of bruxism, such as excessive wear of the bottom incisors, (2) palpation of the head and neck musculature for tender muscles under increased tension, and (3) forceful systematic isometric contraction of muscle groups of the head and neck for their effects upon the patient's tinnitus.

At least three factors have been associated with changes in tinnitus attributes. The first is somatic modulation. As described above it is clear that most, if not all, subjects can somatically modulate their tinnitus. The second is stress. Patients consistently describe that they are more bothered by their tinnitus when stressed. Whether this is due to changes in the tinnitus loudness or because the patient focuses his attention upon the tinnitus frequently can not be distinguished by the patient. In fact, it could be that stress acts through somatic modulation to increase tinnitus loudness, since contractions of cranio-cervical musculature such as clenching the teeth, furrowing the brow, or grimacing often accompany stress. Hence one way by which stress may lead to increased tinnitus loudness is through increasing head and neck muscle contractions which in turn lead to louder tinnitus by the somatic mechanism. Thirdly, some subjects clearly associate an increase in their tinnitus loudness with exposure to loud sound, and in some the louder tinnitus can persist for hours after the exposure has discontinued. Thus, if a patient reports that his tinnitus is intermittent or has wide fluctuations in loudness or other qualities, and there is neither exposure to intense sound nor evidence for stress, then somatic modulation must be suspected.

Table 2.

Tinnitus properties suggesting a somatic component
1. Intermittency
2. Large fluctuations in loudness
3. Variability of location
4. Diurnal pattern
5. No hearing loss but head or neck trauma

A history of variations in tinnitus loudness then raise the suspicion for a somatic factor modulating the percept's loudness (table 2). At an extreme are patients who describe that they have periods when their tinnitus can not be heard, even in the quiet. Others report wide variations in the loudness of their tinnitus. For still others, their tinnitus is unilateral when it is

relatively quiet but becomes non-lateralized when the tinnitus is louder [See case 3]. Such phenomena suggest that there are on-going somatically mediated factors modulating the tinnitus percept.

Diurnal fluctuations in the tinnitus percept also suggest that somatic modulation is operative. Patients who describe their tinnitus as louder upon awakening raise the possibility that somatic factors (such as bruxism - grinding of the teeth) are active during sleep and are causing an increase in tinnitus loudness. Others describe that their tinnitus has usually vanished by the time they awaken and then returns a few hours into the day; this scenario suggests that during the day they are re-activating their tinnitus through somatic mechanisms, such as the tonic muscle contractions required to support the head in an upright position or clenching related to the stress of daily activities. Finally others describe that their tinnitus is louder after awakening from a nap in a chair; this may relate to somatic factors such as stretching of the neck muscles when their head passively falls forward while dozing in a sitting position.

In our experience while a somatic factor on its own can cause tinnitus (Case 1), much more frequently somatic factors combine with other factors (such as chronic hearing loss) to act as trigger factors or modulators (Cases 2 and 3).

Trauma. The contemporaneous association between head trauma, hearing loss and tinnitus makes the diagnosis straightforward. The association with trauma becomes less certain when some of these elements are missing, such as a delay between the trauma and the onset of the tinnitus. The longer the delay the less the confidence in any association. Tinnitus but no hearing loss following trauma, likewise, makes the association more tenuous and raises the possibility that trauma is only indirectly causing tinnitus such as through a somatic mechanism.

Post-infectious: Occasionally patients report the onset of tinnitus following an upper respiratory infection. Whether this is cause and effect has never been established. Since upper respiratory infections are common, and idiopathic tinnitus not uncommon, chance association of the two is to be expected. Our personal observations that tinnitus with upper respiratory infections appears to have its onset several days following the onset of the illness suggests that the association may be more than at a chance level. Such tinnitus has not been accompanied by hearing loss.

Medication-related (including withdrawal syndromes): The temporal association of the onset of the tinnitus with exposure to a toxin establishes the diagnosis particularly if the tinnitus resolves when the toxin is withdrawn. An association is less clear when tinnitus begins just after a new medication was begun but does not remit when discontinued. Such instances raise the possibility that the new medication acted as a trigger factor for the tinnitus. Aside from high dose aspirin and quinine, which can also cause a reversible hearing impairment, the association of tinnitus with medications is anecdotal. Cisplatin, aminoglcoside antibotics, and loop diuretics can cause permanent hearing loss and probably tinnitus. Transient tinnitus can be a part of a sedative withdrawal syndrome.

Central nervous system disorder - rostral to trapezoid body . The evidence available indicates that for central nervous system lesions rostral to the trapezoid body (such as involving the inferior colliculus) tinnitus is usually transient and bilateral (Hausler and Levine 2000) .

Idiopathic. Often, despite an exhaustive evaluation of non-specific tinnitus, no specific diagnosis can be made with any high degree of confidence. Hence, the diagnosis of "idiopathic" is made.

CASE STUDIES: Non-specific tinnitus

CASE 1. Unilateral somatic factor can cause ipsilateral tinnitus.
Ten months prior to her clinic visit, a 29 year old woman developed transient hyperacusis following 36 hours of non-stop work on the telephone. Initially she had "motor noise" in both ears, but within 6 weeks it became "very severe high-pitched right ear tinnitus," that persisted for 6 months. Audiogram and MRI scan were normal. By the time of her evaluation her right ear tinnitus had not been present for 2 months. [She did notice a very faint (1 on a scale of 10) constant ringing of both ears.] On examination her right sternocleidomastoid muscle was tender and under increased tension as compared to the left sternocleidomastoid. Forceful isometric contraction of her right sternocleidomastoid muscle (head turned to left and tilted to the right) elicited right ear tinnitus identical to the tinnitus she had previously experienced for 6 months.

Results of SOMATIC TESTING with 10 brief forceful isometric head and neck contractions:

CONDITION	TINNITUS LOUDNESS	TINNITUS LOCATION
BASELINE (0-10 SCALE)	**1**	**BOTH EARS**
1.EXTENSION	NO CHANGE	
2. FLEXION	**4**	**RIGHT EAR ONLY**
3. CLENCHING TEETH	NO CHANGE	
4. RIGHT LATERAL FLEXION	NO CHANGE	
5. LEFT LATERAL FLEXION	NO CHANGE	
6. TURN RIGHT	NO CHANGE	
7. TURN LEFT	NO CHANGE	
8. VERTEX PRESSURE	NO CHANGE	
9. TURN LEFT, WITH RIGHT LATERAL FLEXION	**4**	**RIGHT EAR ONLY**
10. TURN RIGHT, WITH LEFT LATERAL FLEXION	NO CHANGE	

Comment: The physical findings of a tender, firm right sternocleidomastoid muscle and reactivation of her ipsilateral tinnitus with maneuvers that activate this muscle all point to to her tinnitus being related to this abnormal muscle, possibly through influencing the activity of the dorsal cochlear nucleus (figure 2).

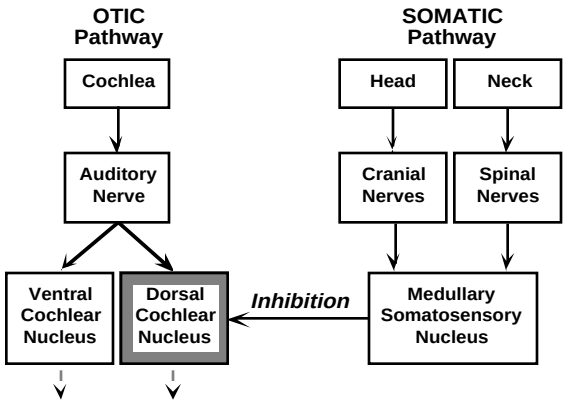

Figure 2. How otic and somatic factors may interact: a neurological model for unilateral tinnitus. Tinnitus may develop due to disinhibition of the dorsal cochlear nucleus via the otic pathway (ipsilateral cochlea or auditory nerve) (Kaltenbach and McCaslin 1996) or via the somatic pathway (ipsilateral head or neck (case 1)) (Levine 1999a) . The way these two inputs influence dorsal cochlear nucleus activity could explain how otic and somatic factors interact to either cause (case 2) or relieve unilateral tinnitus (case 3).

CASE 2. Tinnitus can be multifactorial. Intermittency suggests a somatic component to the tinnitus.

A 50 year old woman presented with a history of a few months of intermittent left ear tinnitus that began when her neck was manipulated. For more than 5 years previously, she had had a unilateral mixed, but predominantly sensorineural left-sided hearing loss due to otosclerosis [mild loss below and moderately severe above 2 kHz]. When initially examined, she had no tinnitus. Her left suboccipital muscles, however, were noted to be under increased muscle tension and more tender to firm pressure than the corresponding muscles on her right side. Within about five minutes of examining these muscles, her left-sided tinnitus began. On re-examination her left suboccipital muscle tension had become much more pronounced. Within another five minutes her tinnitus abated and her suboccipital muscles were again more relaxed.

Comment: Three observations suggest that her unilateral intermittent tinnitus was at least in part somatically mediated. Her tinnitus (1) was said to have begun with neck manipulation, (2) could be brought on by firm pressure applied to her suboccipital musculature, and (3) was closely related to the degree of tension in these muscles. In view of her preexisting predominantly sensorineural ipsilateral hearing loss, it is likely that her tinnitus was multi-factorial: in part due to the longstanding sensorineural hearing loss and in part to the insult to her ipsilateral neck. Neither factor alone may have been adequate to induce tinnitus, but together they interacted to cause her tinnitus (figure 2).

CASE 3. Intermittency, variability in tinnitus location and loudness suggest somatic factors are operative.
An 87 year old woman had a year of intermittent severe hissing tinnitus that could be left ear only or bilateral. It followed a cyclical pattern. Following a day or two of no tinnitus, it would begin softly in the left ear but over the next two days become progressively louder as it spread to both ears. It would then lessen over the next two days or so, until the tinnitus was heard only in the left ear and ultimately would disappear completely. She had a long history of bruxism and muscle contraction headaches (tightness over her temples and jaw). When her tinnitus was severe her headaches were severe. Her audiogram showed a sloping symmetric pattern consistent with her age [thresholds extending from 20 dB at 250 Hz to 70 dB at 8 kHz]. At a visit when her tinnitus was extremely loud (10/10) and bilateral, somatic testing was performed (as in case 1 above). Her tinnitus loudness decreased to 4/10 and became unilateral (left ear only). These changes in her tinnitus persisted for the remainder of that visit (20 minutes). By the next day her tinnitus had disappeared completely, but the usual cycle then returned within a day or two.
Comment. On the one hand, because there was no obvious event that precipitated her tinnitus and her hearing loss was longstanding and chronically progressive, there is no readily apparent cause for her non-specific tinnitus. On the other hand, several points support the diagnosis of somatically related tinnitus: (a) somatic modulation could relieve her tinnitus by "short-circuiting" her cycle, (b) her tinnitus began unilaterally even though her audiogram was symmetric, and (d) her tinnitus cycles tended to parallel her muscle contraction headaches.

CASE 4. Trigger factor.
A 33 year old man was seen because of non-lateralized tinnitus. His mother had been diagnosed with terminal lung cancer about three weeks earlier, and he had been closely involved with her care. Two days before her death as he was trying to sleep, his tinnitus began. It was described as a high-pitched ringing similar in pitch but much louder than transient tinnitus he had previously experienced following loud sound exposure. Clenching or turning his head would aggravate his tinnitus. His exam and audiogram were normal. Despite four years of a variety of treatments, his tinnitus has persisted and he remains distressed.
Comment: No diagnosis could be established with any great confidence to account for his tinnitus. What is striking is the onset of the non-specific tinnitus at an extremely stressful time of his life. The stress of his mother's brief terminal illness and her death appears to have been the "trigger factor" that precipitated his awareness of tinnitus.

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