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Pulsatile tinnitus: a review

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Abstract. *Pulsatile tinnitus: a review*. *Objectives:* The aim of this review is to provide an up to date review of the causes, assessment, and management of pulsatile tinnitus.
Methods: A search was performed of all studies, review articles, and case series looking at pulsatile tinnitus, published until the 1st January 2015. The search was performed using a keyword search strategy (*pulsatile tinnitus, arteriovenous malformation*) on PubMed, Medline/Old Medline and Cochrane Central databases.
Results: Pooling the data from the studies in our literature review, with a total number of patients of 1,245 patients, we found that the most common Aetiologies for pulsatile tinnitus were idiopathic (32%), idiopathic intracranial Hypertension (28%), atherosclerosis of the carotid arteries (16%), Otosclerosis (11%), Glomus tumors (7%), and Arteriovenous malformations (2%). There is no consensus about first-line imaging modality, however, in reviewing the potential causes we suggested that the first line imaging for pulsatile tinnitus should be a delayed Computed Tomography (CT) Angiographic protocol. For the vast majority of pulsatile tinnitus with no identifiable cause, sound therapy is an effective management option if symptoms are persistent and bothersome. Cognitive Behavioural Therapy (CBT) should also be considered if there is significant anxiety or concurrent depressive illness.
Conclusion: It is important to investigate patients with pulsatile tinnitus radiologically to rule out significant causes, and we recommend CT angiography as the first-line imaging modality. Where no cause is found and patients have persistent bothersome symptoms, consideration should be given for treatment with sound therapy and cognitive behavioural therapy.

Introduction

Tinnitus is defined as the perception of sound in one or both ears in the absence of an external stimulus. Pulsatile tinnitus differs in that it is a sound that is heard by the patient, synchronous with their heartbeat. The prevalence of pulsatile tinnitus amongst patients with tinnitus is approximately 4%.[1] It is important to differentiate pulsatile tinnitus from non-pulsatile tinnitus, as in the literature, it was reported that up to 92% of the cases in pulsatile tinnitus that has an underlying demonstrable cause.[2] There is a wide variation given in the literature regarding the incidence of pulsatile tinnitus with an underlying cause, and this is likely to be due to differences in populations, the experience of clinicians, diagnostic beliefs, and reporting mainly from tertiary centers where rare causes are more common. Also, the symptom of pulsatile tinnitus is itself more common than reported, but often not investigated as it may be in conjunction with non-pulsatile tinnitus/deafness or be intermittent/non-bothersome.

Pulsatile tinnitus can be classified as objective or subjective, according to whether it is audible to both the clinician and patient, or to the patient only, respectively. Furthermore, pulsatile tinnitus can be classified as either vascular or nonvascular in origin. When pulsatile tinnitus arises secondary to a vascular cause it is due to turbulent blood flow, either because of increased blood flow or due to stenosis of the blood vessel. This disturbed flow is transmitted directly to the inner ear, causing the sensation of pulsatile tinnitus.[1,2]

The aim of this review is to provide an up to date review of the aetiology, assessment, and management of pulsatile tinnitus. Pulsatile tinnitus was defined as sound that is heard by the patient in one or both ears, synchronous with their heartbeat, in the absence of an external stimulus.

Methods

A search was performed of all review articles and case series looking at pulsatile tinnitus, published until the 1st January 2015. The search was performed

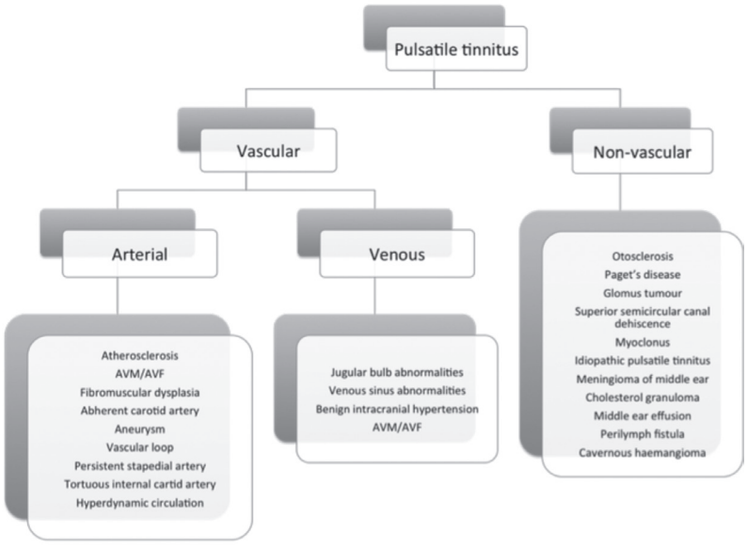

Figure 1
Causes of pulsatile tinnitus.

using a keyword search strategy (*Pulsatile tinnitus, arteriovenous malformation*) on PubMed, Medline/Old Medline and Cochrane Central databases. The reference lists of the manuscripts were also reviewed to identify further studies. To meet the inclusion criteria for the study, studies were required to be full-text articles in English, looking at isolated pulsatile tinnitus. Studies were excluded if there were 5 or less cases reported.

Literature review

The literature review identified 1,793 full-text papers initially. However, only 25 of these papers met our inclusion criteria and were therefore included in our review.[1-25]

Figure 1 summarizes the main causes of pulsatile tinnitus. A study over a 15-year period of 145 patients suggests that the most common causes of pulsatile tinnitus are atherosclerosis of the carotid arteries, idiopathic intracranial Hypertension, and Glomus tumours.[3] In pooling the data from the studies in our literature review, with a total number of 1,245 patients, we found the most common Aetiologies for pulsatile tinnitus were idiopathic (32%), Idiopathic Intracranial Hypertension (28%), atherosclerosis of the carotid arteries (16%), Otosclerosis (11%), Glomus tumours (7%), and Arteriovenous malformations (2%).

Internal carotid artery atherosclerosis is a common cause of pulsatile tinnitus in the population above the age of 50. In a series of 12 patients with pulsatile tinnitus secondary to atherosclerotic disease, all had an ipsilateral carotid bruit.[4] If a bruit is identified, CT angiography is the most appropriate and sensitive imaging modality as opposed to magnetic resonance imaging (MRI).[5]

A recent systematic review has concluded that there is an association between tinnitus and hypertension, but the cause and effect relationship is uncertain.[6]

A dissecting aneurysm of the internal carotid or vertebral artery can manifest as pulsatile tinnitus and is important to recognize. The majority of patients with dissection will be diagnosed using CT angiography, which is the first line imaging modality of choice as it is fast and accurate.

The management of these patients remains controversial – they may be managed with anti-coagulation or they may be managed with Antiplatelet therapy. Past trials have shown no significant difference in the outcome between treating and not treating, but these have involved small cohorts.[7]

Other arterial causes for pulsatile tinnitus include an aberrant internal carotid artery and a persistent stapedial artery, which are usually managed conservatively.[8]

There is a little agreement in the literature over neurovascular compression of the vestibulocochlear nerve causing tinnitus as a result of vascular loops. Some studies have reported Radiological evidence

of vestibulocochlear nerve compression in both asymptomatic and pulsatile tinnitus patients, and have found no significant difference in the spatial position of these vascular loops in relation to the vestibulocochlear nerve in patients with and without pulsatile tinnitus.[9] Cadaveric studies have reported a frequency of vascular loops of up to 12.3%, but this could be affected by the preparation process with formaldehyde.[9] Microvascular decompression is reported to have variable success rates in different papers (27%-100%), and the success is less likely in patients with bilateral tinnitus and/or hearing loss.[10]

Venous causes for pulsatile tinnitus include a high riding jugular bulb (Figure 2) and Diverticula of the Sigmoid and transverse sinuses.

Idiopathic intracranial hypertension (IIH), or benign intracranial hypertension (pseudotumor cerebri syndrome), is one of the most common causes of pulsatile tinnitus. In over 90% of cases, it occurs in obese women of childbearing age. Pulsatile tinnitus may be the only presenting feature, but it can be associated with headaches, blurred vision, low-frequency hearing loss, vertigo, and aural fullness.

Diagnosis is usually confirmed by lumbar puncture and the presence of elevated cerebrospinal fluid (CSF) pressure (>200mm H_2O) with normal CSF constituents.[11] The imaging of choice here to exclude the most common mimic, Dural venous sinus thrombosis, is CT Venography, but Magnetic Resonance Imaging (MRI) can be used as a second line investigation. The majority of the literature suggests that the best treatment is weight loss.[11]

Arteriovenous malformations (AVM) and fistulae (AVF) are a rare cause of pulsatile tinnitus, however, they are often present with an objective pulsatile tinnitus. Dural AVF is the most common AVM. They usually occur between a meningeal artery and vein or venous sinus. CT angiography is the first line and very sensitive (Figure 3).[12] High-flow fistulas usually require surgical treatment in the form of Endovascular Embolization or surgical ligation of the feeding vessel. Radiosurgery has been reported to be successful for larger AVF, with success rates of up to 95%.[12]

Glomus tumours may be present with a reddish mass behind the tympanic membrane. CT angiography or contrast-enhanced MRI can be utilized for assessment of glomus tumours (Figure 4). Glomus tumours on gadolinium-enhanced MR

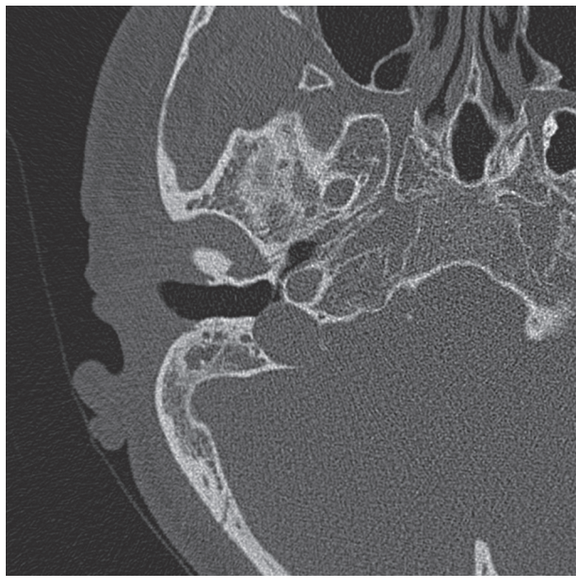

Figure 2
Axial CT image to demonstrate high riding jugular bulb.

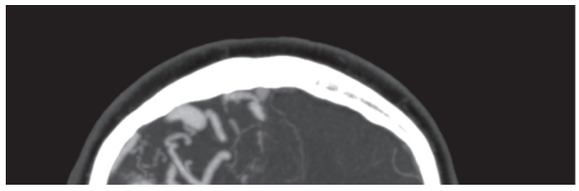

Figure 3
Coronal slice from CT angiogram using delayed CT angiographic protocol, demonstrating ateriovenous malformation.

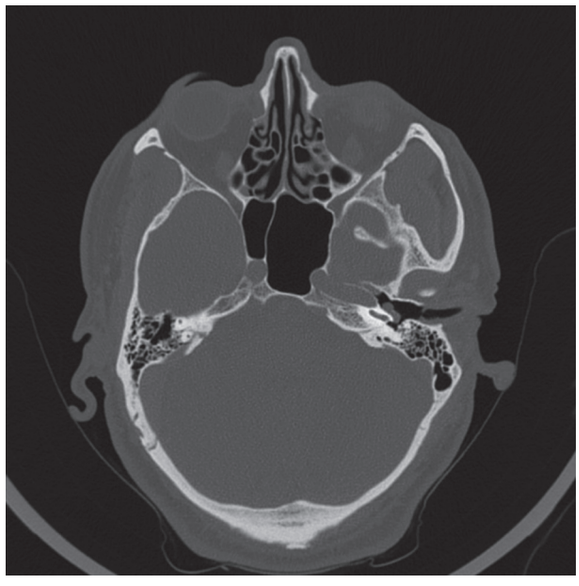

Figure 4
Axial CT image to demonstrate glomus tympanicum

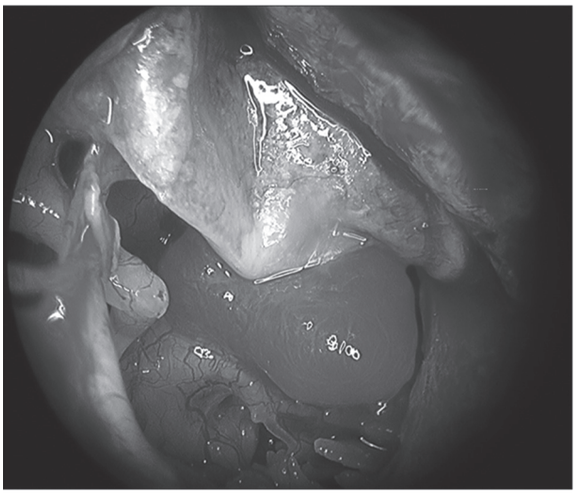

Figure 5
Intraoperative view of glomus tympanicum

imaging have a characteristic 'salt and pepper' appearance.[1] The usual management is surgical resection after pre-operative embolization (Figure 5). Some papers advocate using the Stereotactic Radiosurgery, as it reduced morbidity.[13]

In superior semicircular canal Dehiscence (SSCD), patients may present with Tullio's Phenomenon (caused by the creation of a third window in the vestibular system, in addition to the oval and round window), autophony and pulsatile tinnitus. The pulsatile tinnitus is thought to be due to the increased bone conduction with increased perception of sounds produced by the body itself. Imaging via CT and management is usually surgical (canal occlusion and resurfacing, or obliteration), which has been shown to be very successful.[14]

Myoclonic contractions of the middle ear and the palatal muscles can result in an objective pulsating sound, with contraction frequency ranging from 10-240 contractions/minute, so it can be confused with the arterial pulse, giving rise to the sensation of pulsatile tinnitus. Tympanometry over a prolonged period classically demonstrates a 'sawtooth' pattern. Surgical intervention to divide the middle ear muscles, or botulinum toxin for the palatal muscles, has been shown to be effective.[15]

There are some cases where, despite extensive investigation, no cause can be found for the pulsatile tinnitus, and this is termed idiopathic or essential pulsatile tinnitus. It is thought that this is due to Somatosensory interactions with the central auditory pathway, with the stress/anxiety state, often a significant initiating factor. The theory is that the dorsal cochlear nucleus, which normally suppresses self-generated sounds, is disrupted.

Rare causes of pulsatile tinnitus include temporal bone Meningocele, Pneumocephalus, Perilymph fistulas, cholesterol Granulomas, and cavernous sinus Haemangioma.[3]

Assessment

Pulsatile tinnitus must be evaluated with a thorough history, including asking about the nature of the tinnitus, onset, duration, location, any exacerbating or relieving factors, and distress caused by the tinnitus. Other otological symptoms should be elicited, such as hearing loss, vertigo, and aural fullness. Cardiac risk factors should be assessed. If the tinnitus is of sudden onset, care should be taken to rule out a carotid artery dissection.

Examination with careful otoscopy may pick up any middle ear pathology, such as Glomus tumours, an aberrant carotid artery or high jugular bulb. Tensor tympani myoclonus is rarely associated with rhythmic movements of the tympanic membrane. Assessment of the soft palate with oral and fiberoptic examination will detect any soft palate muscle Myoclonus. Auscultation in the region of the temporal bones, orbits, neck, and chest is essential. In order to determine if pulsatile tinnitus is of venous origin, light digital pressure can be applied to the ipsilateral internal jugular vein (IJV). If this results in cessation of the tinnitus it suggests that the pulsatile tinnitus is of venous origin. Turning the head in the direction of the tinnitus has a similar effect as the Sternocleidomastoid compresses the IJV. The measurement of blood pressure is essential and Fundoscopy must be performed (to check for IIH). A full neurological examination must also be undertaken in the case of IIH to check for any neurological complications. Lumbar puncture and measurement of CSF pressure should be done when IIH is suspected.[1]

Haematological investigations include full blood count, vitamin B12 levels, and thyroid function tests, as Hyperthyroidism and Anaemia can result in a hyperdynamic circulation. Blood glucose, cholesterol levels, and a complete biochemistry profile should be obtained, including alkaline phosphatase (raised levels can be associated with Paget's disease).

Pure tone audiogram and Tympanometry must be performed in patients presenting with pulsatile

Table 1
Conditions that can be picked up by differing imaging modalities

MRI Internal Auditory Meatus/Brain	**CT Angiography**
Tumours (Vestibular schwannoma, meningioma, glomus tumour, middle ear adenoma, arteriovenous malformation)	Carotid artery assessment (atherosclerosis, aberrant carotid artery, aneurysm, dissection, tortuous artery)
Cholesterol granuloma	Arteriovenous fistula
Middle ear effusion	Persistent stapedial artery
	Dural venous sinus assessment (diverticulum, stenosis, stricture)
	Jugular bulb anomaly
	Otosclerosis
	Paget's disease
	Fibromuscular dysplasia of internal carotid artery
	Superior semi-circular canal dehiscence
	Glomus tumour
	Internal auditory canal vascular loop
	Middle ear effusion

tinnitus. If a low-frequency hearing loss is picked up, the audiogram should be repeated while light digital pressure is applied over the IJV. If this results in resolution of the hearing loss, it is pathognomic of venous causes, such as IIH. The Tympanometry can help identify middle ear effusions and masses, such as Glomus tumours.

Imaging is guided by clinical findings. There is a wide difference in the literature with regards to the most appropriate imaging modality.[16] We recommend the use of delayed CT angiography as the primary imaging modality as this will identify arterial as well as potential venous causes (see Table 1 It also incorporates an appropriate high-resolution thin-slice CT interrogation of the skull base for entities such as Glomus Jugulotympanicum or Otosclerosis.

MRI should only be used as second-line imaging if no primary cause has been identified on delayed CT angiography, but there are Sensorineural hearing loss or additional cranial nerve palsies clinically, as the burden of exclusion would be for a small nerve sheath tumour of the vestibular nerves, a cochlear nerve sheath tumour (include an Intracochlear tumour), the less common meningeal Carcinomatosis/Tymphomatosis, or idiopathic Pachymeningeal Hypertrophy. The remainder of the concerning pathologies or low-grade pathologies such as secondary features of idiopathic Intracranial Hypertension can be picked up on the delayed of CT angiography protocol.

Management

If an underlying cause is identified, then management should be directed towards correcting this. Where no cause can be found, if symptoms are not bothersome, explanation and reassurance may be all that is needed. Where symptoms are persistent and bothersome, effective treatment options include sound therapy and cognitive behavioural therapy.[1]

The negative impact that the involuntary perception of sound can induce is symptomatically heterogeneous with patients experiencing frustration, anxiety, depression, insomnia, and concentration difficulties.[17]

Individuals with a significant degree of hearing loss and bothersome tinnitus have a very reasonable prospect of benefiting from an appropriate hearing aid system as a result of the instrument enhancing environmental sound and causing the tinnitus signal to become less salient. Hearing aids are most likely to be of benefit to objective tinnitus sufferers who present with a conductive hearing loss, which serves to enhance the perception of physiological or vascular sounds to the affected ear.[1]

Tinnitus Retraining Therapy (TRT) is another popular audiological intervention that was designed with the goal of enabling patients to habituate to their tinnitus perception. It is derived from the Neurophysiological model for tinnitus, which presupposes that tinnitus-related distress is the result of inappropriate signal processing via

the involvement of auditory perceptual, emotional and reactive systems. The therapy employs the use of directive counselling, relaxation exercises, environmental sound enrichment and wearable noise generators/hearing instruments in accordance with the patient profile.[18] One particular study has revealed total masking to be as clinically efficacious as Tinnitus Retraining Therapy (TRT) for the treatment of subjective percepts over a 12-month period.[19]

Numerous studies have reported CBT as being the most effective evidence-based intervention to enable the amelioration of subjective tinnitus symptoms.[20] The method is applied in order to lessen the psychological distress that manifests as a result of tinnitus perception and does not strive to alter the psychoacoustic parameters of the signal.[21] The core process works by enabling an individual with bothersome tinnitus to formulate a more positive and balanced outlook regarding their circumstance.[22] In practice, CBT for tinnitus is typically applied in conjunction with relaxation exercises, advice on environmental sound enrichment and guided imagery techniques.[23] There is also an evolving evidence base to support the efficacy of combining CBT with mindfulness-based meditation with a number of small-scale pilot studies reporting clinically significant results.[24] It is important to note that there is currently no published evidence to endorse the efficacy of CBT specifically with objective tinnitus.[25]

Conclusion

The most common cause for pulsatile tinnitus is essential pulsatile tinnitus, followed by idiopathic Intracranial Hypertension. It is important to investigate patients with pulsatile tinnitus radiologically to rule out significant causes which are treatable. We recommend CT angiography as the first-line imaging modality for pulsatile tinnitus as most of the significant causes can be diagnosed with the exception of a very small dural fistula. MRI should only be used as second-line imaging if no primary cause has been identified on delayed CT angiography, and there is sensorineural hearing loss or additional cranial nerve palsy clinic. Where no cause is found and patients have persistent bothersome symptoms, consideration should be given for treatment with sound therapy and cognitive behavioural therapy.

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Conflict of interest

None

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