Vestibular Neuritis

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Vestibular neuritis (VN) is the second most common cause of vertigo after BPPV (Neuhauser 2016). It is characterized by a sudden unilateral loss of vestibular function without change in hearing. When hearing loss accompanies the vestibular loss it is referred to as labyrinthitis. The cause of the vestibulopathy remains unclear but possible causes include viral infection, reactivation of herpes family viruses in the vestibular ganglia, vascular causes and possibly autoimmune mechanisms. VN is managed primarily with symptomatic treatments acutely and vestibular physical therapy.

Vestibular neuritis synonyms: vestibular neuronitis, labyrinthitis, neurolabyrinthitis, acute vestibular paralysis, epidemic vertigo

Acute Vestibular Syndrome (AVS) – acute vertigo characterized by (Hotson 1998, Venhovens 2016)

1. Acute onset of dizziness/vertigo with nausea/ vomiting
2. Worsened vertigo with head motion
3. Spontaneous or gaze induced nystagmus
4. Gait unsteadiness
5. Duration of 24 hours up to several weeks.

Epidemiology

- Incidence ranges between 11.7 to 15.5 per 100,000 per year and mean age is about 52 with a female: male ratio of 1.1:1.0 and no seasonal clustering (Adamec 2015)
- Vestibular neuritis is more common than labyrinthitis. Labyrinthitis is more likely to show damage of vestibular structures innervated by the inferior vestibular nerve (Pogson 2016).
- About 5.6% of cases presenting with acute vertigo in an ED are due to vestibular neuritis or labyrinthitis (Kim 2008)
- VN may account for 3-10% of dizziness patients in a specialty clinic population and is second in frequency to BPPV (Neuhauser 2016)

Etiology/Pathophysiology

The cause of VN is still unknown. Proposed theories of causation include reactivation of latent HSV-1 or occasionally VZV, direct viral infections, vascular occlusion, and immune mediated mechanisms (Greco 2014). Some and perhaps many cases are attributable to reactivation of latent HSV in the vestibular ganglia that become reactivated causing the syndrome (Arbusow 2010).

There is a predilection for the superior vestibular nerve to be affected which may be due to the narrower canal traversed by the superior versus the inferior vestibular nerve.

Common co-morbid conditions hypertension 30.4%) DM 8.9%), dyslipidemia (7.5%), hypothyroidism (6.3%). Fewer than half of patients had a patients with VN had prior viral illness (Norrving 1995).

Clinical History

Case History:

46 year old man had the onset of vertigo at 7 am. He felt some dizziness turning in bed and when he got out of bed he staggered and had to hold on to walk to the bathroom. The dizziness continued to worsen over the subsequent 45 minutes and felt like spinning with nausea and vomited with dry heaves. He noted spinning even holding still but all the symptoms worsened with

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any kind of head movement. He presented to the ED 2 hours after the onset due to persisting vertigo and nausea.

Onset usually evolves over 20 min to several hours. Initial dizziness may be described as something other than spinning. Occasionally there is a bit of a stuttering onset with some dizziness or vertigo, a limited symptom-free interval for days to as long as a week, then full onset. The degree of nausea and vomiting depends on the severity, the parts of the labyrinth most affected and also on the individual's innate susceptibility to nausea, which may vary. Patients learn to lie still and avoid head movement as this worsens the vertigo.

Examination

Case History (cont'd):

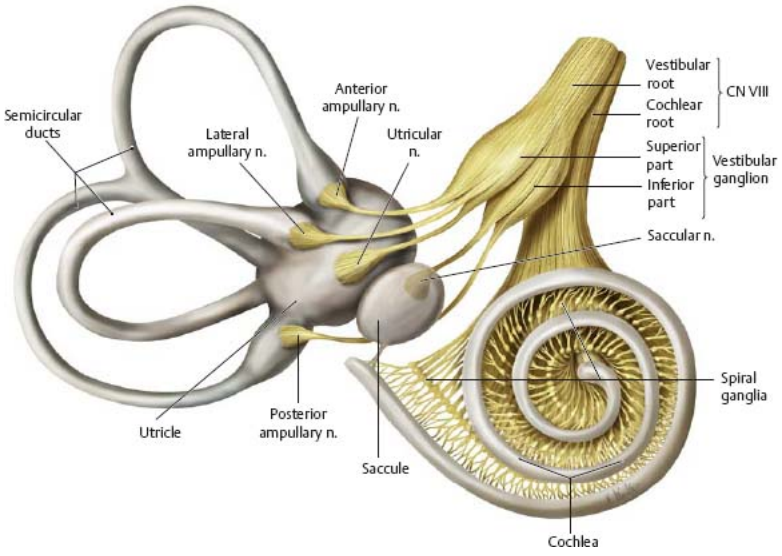
On exam he had mild unsteadiness in walking. Speech was clear. Finger-nose coordination was intact. There was spontaneous right beating nystagmus intensified when looking to the right and lessened by looking to the left. The remainder of the neurological exam was normal and hearing was intact bilaterally to bedside testing. Gait was reluctant and cautious but he was able to walk with hands outstretched in a guarded manner.

Patients may appear sweaty or ashen if nausea is intense. Speech articulation and limb coordination are intact. Gait notable for some degree of imbalance with some tendency to fall toward the affected side. Fukuda test tends to cause deviation toward the affected side. Past pointing may cause deviation toward the side of the affected ear.

Nystagmus occurs due to vestibular asymmetry and the fast phase beats *away* from the affected ear. Compensation and thus abatement of nystagmus varies but is often attenuated within 24-48 hours but can be observed often for weeks afterwards. Initially the nystagmus is spontaneous, then improves so it is only seen with gaze to the side of the unaffected ear. There is often static positional nystagmus seen even after the spontaneous and gaze nystagmus are no longer present. Caloric asymmetry and head impulse test abnormalities persist, provided there is truly substantial permanent vestibular hypofunction.

Types of vestibular neuritis

- Superior vestibular nerve supplies: anterior canal, horizontal canal, utricle
- Inferior vestibular nerve supplies: posterior canal, saccule
- Complete vestibular neurits = both superior and inferior nerve distribution affected.



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Available vestibular tests:

- Anterior, horizontal, posterior semicircular canal function – **video head impulse testing (vHIT)**
- Horizontal canal function – **head impulse (bedside), caloric testing (as done with VNG)**
- Utricle – **ocular vestibular evoked myogenic potentials (oVEMP)**
- Saccule – **cervical vestibular evoked myogenic potentials (cVEMP)**

Differential Diagnosis

- Meniere's, first attack – Meniere's disease is characterized by severe vertigo usually lasting 20 min to 12 hours (Lopez-Escamez 2015) whereas VN recurrences are very uncommon and vertigo lasts > 24 hours and usually for several weeks.
- Vestibular migraine (VM) – VM can have a number of differing profiles of vertigo ranging from chronic motion intolerance to brief vertigo attacks lasting minutes. Some cases of VM can cause attacks of vertigo with nausea and vomiting and without hearing change. VN is usually a single attack, monophasic and lasts > 24 hours with gradual recovery and characteristic nystagmus not seen with VM.
- Cerebellar stroke – Strokes presenting with isolated prolonged vertigo can mimic VN and will be discussed more below. In a study of 114 AVS admitted to a general hospital, 72/114 had VN, 42/114 had a stroke (32/114 PICA, 10/114 AICA (Lee 2009). The ocular motor exam is very helpful in distinguishing VN from stroke as the cause of prolonged isolated vertigo (Newman-Toker 2013).
- Less similar but sometimes misdiagnosed: BPPV, otitis media, food poisoning

	Inferior cerebellar infarct	Vestibular neuritis
Nystagmus	Spontaneous nystagmus and gaze-evoked nystagmus (change of direction of nystagmus with change of direction of gaze). Nystagmus may be only with gaze in one direction. Vertical spontaneous nystagmus.	Spontaneous and Direction-fixed nystagmus strongest with when looking in the direction of the fast phase of nystagmus. Nystagmus is lessened with visual fixation.
Accompanying signs	Diplopia, slurred speech, limb dysmetria, Horner's syndrome, unilateral sensory loss	Absent; ipsilateral acute hearing loss in the case of labyrinthitis.
Severity of postural imbalance	More severe or unable to walk (Lee 2009)	Often can walk with some effort
Hearing loss	None*	Ipsilateral hearing loss in 10-20%
Head Impulse test	Normal	Abnormal

* Exception is lateral pontine infarction due to AICA branch occlusion in which hearing is lost due to labyrinthine infarction or infarct of the cochlear nerve root entry zone and this may be combined with cerebellar infarction but often there is also some unilateral facial paresis and ipsilateral facial hypesthesia and possibly ipsilateral Horner's syndrome (Lee 2009, Kim 2017).

Acute audiovestibular loss may sometimes be a sign of an impending AICA territory infarction due to occlusive vascular disease of the vertebral or basilar artery near the origin of AICA (Kim 2017). There is no widely accepted algorithm to identify the cause of isolated prolonged vertigo due to stroke versus VN.

The odds of stroke go up with co-morbidities of CAD, DLP and HTN (Chase 2014). Perhaps the best method so far is "HINTS". This acronym stands for Head Impulse, Nystagmus, and Test of Skew. The sensitivity of HINTS for identifying stroke was 96.5% (109/113) and the specificity was 84.4% (65/77)(Newman-Toker 2013).

A study will soon be exploring whether a computer-based HINTS algorithm based on video-goggle findings can be used in an ED setting to aid in deciding whether a stroke or peripheral vestibular cause is more likely. HINTS is limited now because it requires expert interpretation. Furthermore, HINTS has yet

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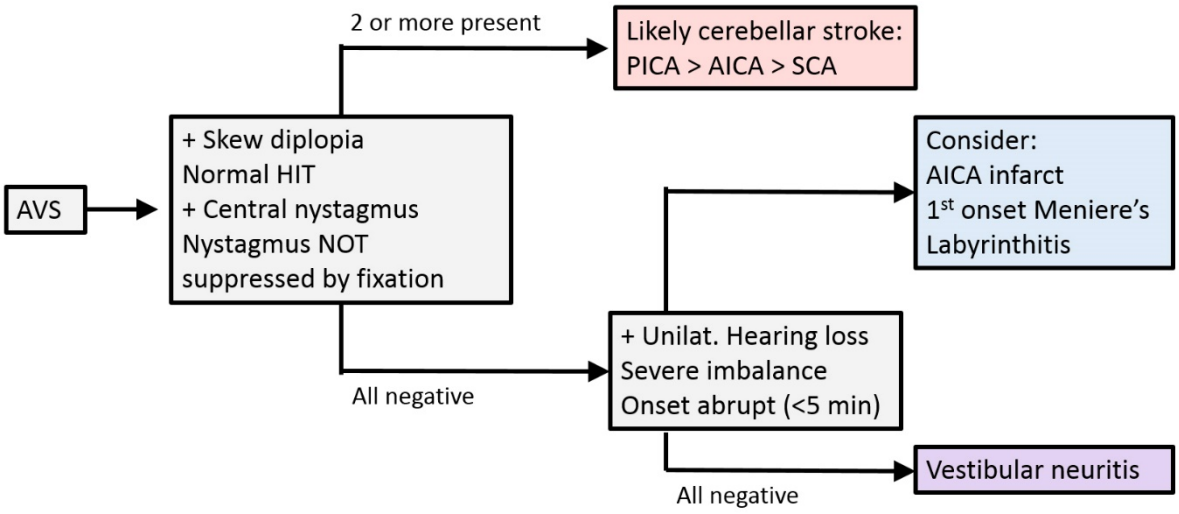
to be validated and pre-determined cut-off points between normal and abnormal findings must be determined and standardized (Newman-Toker 2017).



Adapted from Venhovens J et al. J Neurol 2016;263(11):2151-2157.

4-11% of strokes may present with isolated vertigo and be misdiagnosed as vestibular neuritis (Venhoven 2016, Norrving 1995). Fewer than half of patients had a patients with VN had prior viral illness (Norrving 1995).

Prognosis

- In a study of 40 VN patients with >25% caloric asymmetry were studied at the onset and then again at 6 weeks (Kim HA 2008):
  - 100% had caloric vestibular loss (>25% asymmetry) at onset but only 78% at 6 weeks
  - 100% had vertigo at outset and none had true vertigo at 6 weeks.
  - 100% had abnormal HIT at onset and only 45% had HIT abnormality at 6 weeks
- Another study found 50% recovery of canal function (Buki 2016)
- Recurrence rate is about 2% (Neuhauser 2016)
- Persistent symptoms of dizziness occurred in 30-40% of VN patients, in part to interval development of anxiety or vestibular migraine (Neuhauser 2016) but a small number of patients appear to have especially poor compensation.

Treatment

- Corticosteroids improves the degree of caloric asymmetry but has not been shown to improve function or symptom outcome (Fishman 2011).
- Valacyclovir alone or in combination with corticosteroids for all comers with VN was not helpful (Strupp 2004)
- Vestibular physical therapy improves outcomes in patients with unilateral vestibulopathy, according to two different evidence-based reviews (Hillier 2011, Hall 2016).

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