

Acute unilateral vestibulopathy/vestibular neuritis: Diagnostic criteria

Consensus document of the committee for the classification of vestibular disorders of the Bárány Society

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Received 26 February 2022

Accepted 24 May 2022

Abstract. This paper describes the diagnostic criteria for Acute Unilateral Vestibulopathy (AUVP), a synonym for vestibular neuritis, as defined by the Committee for the Classification of Vestibular Disorders of the Bárány Society. AUVP manifests as an acute vestibular syndrome due to an acute unilateral loss of peripheral vestibular function without evidence for acute central or acute audiological symptoms or signs. This implies that the diagnosis of AUVP is based on the patient history, bedside examination, and, if necessary, laboratory evaluation. The leading symptom is an acute or rarely subacute onset of spinning or non-spinning vertigo with unsteadiness, nausea/vomiting and/or oscillopsia. A leading clinical sign is a spontaneous peripheral vestibular nystagmus, which is direction-fixed and enhanced by removal of visual fixation with a trajectory appropriate to the semicircular canal afferents involved (generally horizontal-torsional). The diagnostic criteria were classified by the committee for four categories: 1. “Acute Unilateral Vestibulopathy”, 2. “Acute Unilateral Vestibulopathy in Evolution”, 3. “Probable Acute Unilateral Vestibulopathy” and 4. “History of Acute Unilateral Vestibulopathy”. The specific diagnostic criteria for these are as follows:

“Acute Unilateral Vestibulopathy”: A) Acute or subacute onset of sustained spinning or non-spinning vertigo (i.e., an acute vestibular syndrome) of moderate to severe intensity with symptoms lasting for at least 24 hours. B) Spontaneous peripheral vestibular nystagmus with a trajectory appropriate to the semicircular canal afferents involved, generally horizontal-torsional,

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direction-fixed, and enhanced by removal of visual fixation. C) Unambiguous evidence of reduced VOR function on the side opposite the direction of the fast phase of the spontaneous nystagmus. D) No evidence for acute central neurological, otological or audiological symptoms. E) No acute central neurological signs, namely no central ocular motor or central vestibular signs, in particular no pronounced skew deviation, no gaze-evoked nystagmus, and no acute audiological or otological signs. F) Not better accounted for by another disease or disorder.

“Acute Unilateral Vestibulopathy in Evolution”: A) Acute or subacute onset of sustained spinning or non-spinning vertigo with continuous symptoms for more than 3 hours, but not yet lasting for at least 24 h hours, when patient is seen; B) - F) as above. This category is useful for diagnostic reasons to differentiate from acute central vestibular syndromes, to initiate specific treatments, and for research to include patients in clinical studies.

“Probable Acute Unilateral Vestibulopathy”: Identical to AUVP except that the unilateral VOR deficit is not clearly observed or documented.

“History of acute unilateral vestibulopathy”: A) History of acute or subacute onset of vertigo lasting at least 24 hours and slowly decreasing in intensity. B) No history of simultaneous acute audiological or central neurological symptoms. C) Unambiguous evidence of unilaterally reduced VOR function. D) No history of simultaneous acute central neurological signs, namely no central ocular motor or central vestibular signs and no acute audiological or otological signs. E) Not better accounted for by another disease or disorder. This category allows a diagnosis in patients presenting with a unilateral peripheral vestibular deficit and a *history* of an acute vestibular syndrome who are examined well after the acute phase.

It is important to note that there is no definite test for AUVP. Therefore, its diagnosis requires the exclusion of central lesions as well as a variety of other peripheral vestibular disorders. Finally, this consensus paper will discuss other aspects of AUVP such as etiology, pathophysiology and laboratory examinations if they are directly relevant to the classification criteria.

Keywords: Acute unilateral vestibulopathy, vestibular neuritis, vestibular neuronitis, acute vestibular syndrome, international classification of vestibular disorders, b \acute{a} r \acute{a} ny society

Synonyms: Vestibular neuronitis, acute vestibular loss, acute vestibular failure

1. Introduction

Acute Unilateral Vestibulopathy (AUVP), also called vestibular neuritis, is an acute peripheral vestibular syndrome defined by an acute unilateral loss of peripheral vestibular function without evidence for acute central neurological or acute audiological symptoms or signs.

The B \acute{a} r \acute{a} ny Society, representing the international community of basic scientists, otolaryngologists, neurologists, audiologists, and therapists committed to vestibular research formed a Classification Committee for an International Classification of Vestibular Disorders (ICVD). Individual disorders are defined by subcommittees that include clinicians and scientists from at least three continents. Since the beginning of the process, the following consensus papers have already been published (<https://www.jvr-web.org/ICVD.html>): Classification of Vestibular Symptoms [17], Vestibular Migraine [95], Meni \acute{e} re’s Disease [98], Benign Paroxysmal Positional Vertigo [154], Vestibular Paroxysmia [147], Persistent Postural-Perceptual Dizziness [141], Bilateral Vestibulopathy [146], Classification of Vestibular Signs and Examination techniques: Nystagmus and

Nystagmus-like Movements [45], Hemodynamic Orthostatic Dizziness/Vertigo [78], Presbyvestibulopathy [2], Mal de D \acute{e} barquement Syndrome [27], Vestibular Migraine of Childhood and Recurrent Vertigo of Childhood [152], Superior Semicircular Canal Dehiscence Syndrome [157], and Motion Sickness [28].

1.1. Terminology and its history

The symptoms and signs of acute vestibular dysfunction have been known for more than 100 years with various terms used. Evidence of the first description of AUVP in the literature – with all limitations of searching due to different languages - is dated from January 27th, 1908. At the Annual meeting of the Austrian Otological Society, B. Rutin precisely and accurately described a case of AUVP with typical symptoms, signs, and findings in caloric irrigation. Since he presumed that it was caused by inflammation, he called it “Neuritis vestibularis” [132]. In 1924 Nylen used the analogous term “vestibular neuritis,” [121] and Hallpike, in 1949, used the term “vestibular neuronitis” [62]. All of these terms implied an inflammation of the vestibular nerve as the etiology of the

disorder, which was supported by histopathological studies (see below). Later, more neutral terms were used, e.g., “unilateral sudden partial loss of vestibular function” by Hemenway and Lindsay in 1956 [97], “vestibular neuropathy” by Haas and Becker in 1958 [60], “vestibuloneuropathy” by Drachman and Hart in 1972 [42], “acute vestibulopathy” by Rau in 1974 [127], “vestibular failure” and “acute peripheral vestibulopathy” by Hess and Reisine in 1984 [67] and “acute unilateral vestibulopathy” by Fetter and Dichgans in 1996 [146].

2. Methods

This work forms part of an ongoing multi-year project to develop an International Classification of Vestibular Disorders (ICVD) that uses a structured process to develop international consensus definitions for vestibular symptoms, syndromes, disorders, and diseases [17]. This process, overseen by the Classification Committee of the Bárány Society (CCBS), is based on expert, multi-disciplinary committees with international representation developing diagnostic criteria for subsequent comment and refinement prior to publication. Thereafter, the criteria are open for comments by the members of the Bárány Society. These criteria are based on a critical appraisal of current best scientific evidence. All definitions are supported by notes, comments, and written discussion according to a template established by the CCBS for ICVD [17]. The criteria for AUVP were developed through discussion, presentation, and refinement from 2015 to 2021. Special care was taken to assure the criteria were practical and applicable in most medical practices.

3. Diagnostic criteria

The committee preferred to use the neutral term “Acute Unilateral Vestibulopathy” (AUVP) [50] (analogous to the term “Bilateral Vestibulopathy,” for internal consistency). The term “vestibular neuritis” can also be used synonymously since it has been the most commonly used term. Inflammation, namely due to the reactivation of a latent herpes simplex - 1 (HSV-1) infection (see below) is the most likely cause but other etiologies are possible.

Three closely related entities were also defined: “AUVP in evolution” a category useful both for practical reasons in the very acute phase to differentiate

from other acute central vestibular syndromes, to initiate specific treatments, or to include patients in clinical studies; “Probable AUVP” if the unilateral peripheral vestibular deficit is uncertain; and “History of AUVP” for the diagnosis of patients who are seen long after the acute phase.

3.1. Diagnostic criteria for “Acute Unilateral Vestibulopathy”

Each of the following criteria have to be fulfilled:

- A) Acute or subacute onset¹ of sustained² spinning or non-spinning vertigo³ (i.e., an acute vestibular syndrome) of moderate to severe intensity⁴ with symptoms lasting for at least 24 hours^{5,6}
- B) Spontaneous peripheral vestibular nystagmus⁷ i.e., a nystagmus with a trajectory appropriate to the semicircular canal afferents involved, generally horizontal-torsional, direction-fixed⁷, and enhanced by removal of visual fixation^{7,8}.
- C) Unambiguous evidence of reduced VOR function^{10,11,12} on the side opposite the direction of the fast phase of the spontaneous nystagmus
- D) No evidence for acute central neurological symptoms or acute audiological symptoms such as hearing loss or tinnitus¹³ or other otologic symptoms such as otalgia
- E) No acute central neurological signs, namely no central ocular motor or central vestibular signs¹⁴, in particular, no skew deviation¹⁵, no gaze-evoked nystagmus¹⁴, and no acute audiological signs¹⁶
- F) Not better accounted for by another disease or disorder

3.2. Diagnostic criteria for “Acute Unilateral Vestibulopathy in Evolution”

Each of the following criteria have to be fulfilled:

- A) Acute or subacute onset¹ of sustained² spinning or non-spinning vertigo³ (i.e., the acute vestibular syndrome) of moderate to severe⁴ intensity, with continuous symptoms for more than 3 hours, but that have not yet lasted for at least 24 hours⁶
- B) Spontaneous peripheral vestibular nystagmus⁷ which is direction-fixed⁷ and enhanced by removal of visual fixation^{7,8} with a trajectory

appropriate to the semicircular canal afferents involved (generally horizontal-torsional)⁹

- C) Unambiguous evidence of reduced VOR function^{10,11,12} on the side opposite the direction of the fast phase of the spontaneous nystagmus.
- D) No evidence for acute central neurological symptoms or acute audiological symptoms such as hearing loss or tinnitus¹³ or other otologic symptoms such as otalgia
- E) No acute central neurological signs, namely no central ocular motor or central vestibular signs¹⁴, in particular, no skew deviation¹⁵, or gaze-evoked nystagmus¹⁴, or acute audiological signs.¹⁶
- F) Not better accounted for by another disease or disorder.

3.3. Diagnostic criteria for “Probable Acute Unilateral Vestibulopathy”

- A) Acute or subacute onset¹ of sustained² spinning or non-spinning vertigo³ (i.e., an acute vestibular syndrome) of moderate to severe intensity⁴ with symptoms lasting for at least 24 hours⁵.
- B) Spontaneous peripheral vestibular nystagmus⁷ which is direction-fixed⁷ and enhanced by removal of visual fixation^{7,8} with a trajectory appropriate to the semicircular canal afferents involved (generally horizontal-torsional)⁹
- C) No clear evidence of reduced VOR function by bedside examination^{10,11,12} on the side opposite the direction of the fast phase of the spontaneous nystagmus.
- D) No evidence for acute central neurological symptoms or acute audiological symptoms such as hearing loss or tinnitus¹³.
- E) No acute central neurological signs, namely no central ocular motor or central vestibular signs¹⁴, in particular, no skew deviation¹⁵, no gaze-evoked nystagmus¹⁴, and no acute audiological signs.¹⁶
- F) Not better accounted for by another disease or disorder.

3.4. Diagnostic criteria for “History of acute unilateral vestibulopathy”

- A) History of acute or subacute¹ onset of sustained² spinning or non-spinning vertigo³

lasting at least 24 hours (i.e., an acute vestibular syndrome) and slowly decreasing in intensity over days.

- B) No history of simultaneous acute central neurological or audiological symptoms such as hearing loss or tinnitus¹³.
- C) Evidence of unilaterally reduced VOR function^{10,11,12,17}.
- D) No history of simultaneous acute central neurological or audiological signs¹³.
- E) Not better accounted for by another disease or disorder.

3.4.1. Notes

1. Up to 25% of patients may experience an episode of vertigo or dizziness lasting for several hours within two days before the onset of the sustained vertigo [87]. If there was an episode of vertigo more than many days before an episode of sustained vertigo, other differential diagnoses should be considered, namely Menière’s disease, vestibular migraine, recurrent vestibulopathy, or TIA (see Comment 4.6., differential diagnosis).
2. Spinning or non-spinning vertigo [17] is continuous, persisting at rest, and usually exacerbated by any head or body movement.
3. Patients often typically complain of vertigo, head motion intolerance, oscillopsia, postural imbalance with a tendency to fall toward the presumably affected side and/or nausea/vomiting.
4. Moderate intensity means that basic activities such as walking a short distance are possible. Severe intensity means that patients are very sick and bedbound. Treatment with anti-vertiginous drugs may reduce symptom severity.
5. A treatment with anti-vertiginous drugs and/or steroids may reduce the intensity and duration of signs (e.g., nystagmus) and symptoms of AUVP.
6. If the acute vestibular syndrome fully recovers before 24 h of duration, the diagnosis of AUVP is unlikely and other differential diagnoses should be considered (see Comments 4.6).
7. Spontaneous peripheral vestibular nystagmus should have the following characteristics: 1) binocular and conjugate; 2) beats in a plane and direction in head-referenced coordinates, regardless of gaze position; in AUVP it is

- typically horizontal-torsional; 3) obeys Alexander's law [128], i.e., an increase of intensity when looking in the direction of the fast phase; 4) reduced by visual fixation and enhanced by lack of fixation, and 5) constant-velocity slow phases if recorded by oculographic equipment. See also "Classification of vestibular signs and examination techniques: nystagmus and nystagmus-like movements" [45].
8. An examination is necessary with Frenzel's glasses [52], similar devices, or a video camera system. A horizontal-torsional nystagmus, not reduced by visual fixation, should not be considered to be of peripheral vestibular origin [66].
 9. Since the horizontal and anterior semicircular canal afferents ("superior vestibular neuritis", see Comments 4.7) are most often impaired, a horizontal-torsional nystagmus with a small upward component is observed. In rare "inferior vestibular neuritis" a vertical-torsional nystagmus is observed (see Comments 4.3.1).
 10. "Unambiguous evidence" refers to a quantitatively demonstrable deficit, such as impaired unilateral VOR gain on the video head impulse test (vHIT) or a unilateral reduced vestibular response on caloric irrigation. The members of the committee are aware of the low sensitivity and specificity of the bedside HIT [102, 162]. However, clinician-assessed refixation saccades during non-quantitative bedside HIT can also be considered as "unambiguous evidence" of an impaired VOR gain when an experienced examiner sees refixation saccades of large amplitude and clearly distinct from the nystagmus beats.
 11. The members of the committee strongly recommend a vHIT because of its high value for the diagnosis of AUVP [109] and because vHIT can be used in the emergency department for quantitative testing and for discrimination between peripheral and central lesions [31, 106, 133]; if the vHIT is normal, caloric testing may be indicated.
 12. The members of the committee are aware that there is so far no general agreement on the pathological cut-off values for results from vHIT or caloric testing for the diagnosis of AUVP [144]. Calculations may depend upon the equipment and analyzing system used. Thus, investigations have to rely on laboratory standards or manufacturers' data. However, a working approximation would be to consider a reduction of gain < 0.7 for vHIT and or a side difference of > 0.3 and/or a caloric side difference of $> 25\%$ as pathological. Furthermore, one should be aware of possible age-dependent decreases in vestibular function (see "Presbyvestibulopathy" [2]).
 13. This requires a systematic patient history with explicit questions about these symptoms, such as hemianopia, double vision (which can also be caused by a decompensation of a strabismus during AUVP), impaired sensation in the face, body, or limbs, problems speaking or swallowing, weakness of the face or limbs, particularly unilateral, and impaired coordination. If a patient develops such symptoms during the course of the disease, the diagnosis has to be reevaluated (see Comment 4.6). New onset of hearing loss, clinical signs, tinnitus or ear pain is not compatible with the diagnosis of AUVP; if attacks re-occur, Menière's disease, vestibular migraine, recurrent vestibulopathy, or TIA should be considered (see Comments 4.6., differential diagnosis).
 14. This requires a systematic neurological examination, namely (a) a neuro-ophthalmological examination that includes the following information: type of nystagmus, eye position with cover tests, range of eye movements, gaze-holding function, smooth pursuit, and saccades [36, 75], (b) an examination of the cranial nerves, e.g., looking for hemianopia, impaired sensation in the face, facial weakness, impairment of hearing, changes in the external auditory canal, dysarthrophonia or problems swallowing, (c) reduced sensation, weakness or impaired coordination of the limbs, i.e., hemihypesthesia, -paresis-, hemiataxia, and (d) unilateral deficits of stance and gait. This implies that the diagnosis of AUVP is a diagnosis after exclusion [148] of an acute central vestibular syndrome due to a brainstem or cerebellar lesion.
 15. A small skew deviation (SD) is present in some patients with an acute unilateral peripheral vestibular lesion (presumably due to damage of utriculo-ocular afferents) [4, 46, 59, 134]. However, a prominent skew deviation is substantially more common among patients with an acute central vestibular syndrome (ACVS), such as stroke, particularly those involving the lateral medulla or pons. As such, the presence of an observable SD using cover and alternating

cover tests at the bedside contravenes a strong diagnosis of AUVP. A study with quantitative measurement of SD showed that a small SD ($<3^\circ$) is seen in about 20% of patients with AUVP [84], so a small SD ($<3^\circ$) should not be used to exclude an AUVP diagnosis; in ACVS, about 30% of patients had a SD and a prominent SD ($>3.3^\circ$) suggested a central lesion [84].

16. If a patient develops central neurological or otological signs such as ear pain during the course of the disorder, the diagnosis has to be re-evaluated.
17. This requires a systematic patient history with explicit questions about these symptoms as well as an evaluation of previous reports with particular attention to such symptoms and signs (see Comments 4.6., differential diagnosis).
18. Some patients with a history consistent with an AUVP have normal vestibular testing at the time of examination. They might have had an AUVP that has recovered but other differential diagnoses could present similarly (see Comments 4.6., differential diagnosis). Therefore, the category of History of AUVP can be applied only if a unilateral lesion is present and other causes of the unilateral vestibular loss are deemed unlikely. Clinicians should be aware that the proposed criteria for unilateral vestibular hypofunction will not give any information regarding the onset of this lesion. A directional preponderance of nystagmus on caloric or rotational testing or the presence of spontaneous vestibular nystagmus are hints that the onset of unilateral hypofunction is recent. However, these abnormalities have low specificity [64].

4. Comments

4.1. Epidemiology

Since the diagnostic criteria for AUVP/vestibular neuritis have not been uniform to date, there are so far no valid state-of-the-art epidemiological studies on AUVP [118]. An annual incidence of 3.5 to 15.5 per 100,000 persons was reported [1, 138]. In a large cohort of more than 36,000 patients who had a standardized evaluation, AUVP was the sixth most common cause of vertigo/dizziness and the third most common cause of peripheral vestibular disorders (BPPV ranks first, Menière's disease second)

[143]. This is also true for children [56, 73]. The usual age of onset is between 30 and 60 years [1, 39], with age distribution plateauing between 40 and 50 years [1, 138]. There is no significant gender difference. Two studies found no evidence for seasonal differences [1, 83]. The reported recurrence rate varies between 1.9% [72] and 10.7% [81].

Patients with AUVP can suffer from subsequent "post-infectious" BPPV of the posterior semicircular canal: in one study 13 out of 104 patients with BPPV had a history of AUVP [23] presumably due to the combination of sparing of the inferior vestibular nerve (see below) and indirect evidence that there is also an inflammation of the labyrinth [8], which increases the risk of a dislodgement of otoconia. AUVP is the third most common trigger after BPPV and vestibular migraine of secondary functional dizziness: in a cohort of 162 patients with secondary functional dizziness, 25 had prior AUVP [61].

4.2. Patient history

The most common symptoms of AUVP are as follows: a) Acute or rarely, subacute, onset of sustained spinning vertigo, with symptoms untreated lasting at least 24 hours. There are no antecedent signs or triggers, except for occasional spells of episodic vertigo a few days before the onset of sustained vertigo in some patients [87]. The vestibular symptoms typically worsen during head and body movements, so patients intuitively try to avoid any movements. b) Apparent movement of the visual surroundings due to a spontaneous nystagmus. c) Postural imbalance with a tendency to veer or fall in the direction of the slow phase of nystagmus, i.e. toward the side of the affected ear. d) Nausea and often vomiting.

To establish a diagnosis of AUVP, the absence of a coinciding acute hearing loss, tinnitus, ear pain, or neurological symptoms, in particular from the brainstem and cerebellum, is required. Patients should be asked about symptoms such as double vision, problems speaking or swallowing, impaired sensation or weakness of the face or body, or other events such as recent trauma.

4.3. Bedside examination

The following bedside tests are relevant for the diagnosis of AUVP and the exclusion of other disorders in the differential diagnosis.

4.3.1. Examination for nystagmus with and without Frenzel's goggles or similar devices that reduce visual fixation

In AUVP there is a spontaneous peripheral vestibular nystagmus that is typically reduced in amplitude by visual fixation due to fixation suppression of the VOR [66, 107]. However, a severely nauseated patient may not be able to cooperate adequately during the acute presentation of symptoms and in the very acute stage of AUVP patients often are not able to significantly suppress the nystagmus. Typically, the intensity of spontaneous nystagmus is enhanced with Frenzel's goggles or similar devices. Eyelid closure can also increase nystagmus, which can be observed or palpated through closed eyelids. A nystagmus that is not reduced in intensity by visual fixation is not a peripheral vestibular spontaneous nystagmus. Some central types of spontaneous nystagmus, e.g., in brainstem infarction, can be reduced by fixation. Therefore, the presence of fixation suppression does not rule out a central lesion. The magnitude of suppression is lower in patients with central nervous system abnormalities compared to patients with peripheral vestibular abnormalities [107].

According to Alexander's law, nystagmus amplitude and slow-phase velocity are increased with gaze toward the direction of the fast phase and decreased with gaze toward the direction of the slow phase of the nystagmus. The nystagmus does not change direction.

The direction of nystagmus corresponds to the semicircular canal afferents involved. The vast majority of patients with AUVP have a "superior AUVP", i.e. an impairment of the function of the horizontal and anterior semicircular canals. There is a horizontal-torsional nystagmus with the fast phase beating toward the non-affected ear with a torsional component such that the upper pole of the eye beats in the direction of the fast phase of the horizontal component. There is often a small upward component [50, 159].

A pure vertical, pure horizontal, or pure torsional nystagmus also is not compatible with a diagnosis of AUVP (for Ref. see [45, 94]).

In "inferior AUVP" [20, 116] the direction of the spontaneous nystagmus corresponds to the plane of the posterior semicircular canal, with a torsional and downward direction. [80]. Inferior AUVP is often misdiagnosed as "central".

If the superior and the inferior vestibular nerves are both affected ("total AUVP"), a pure horizontal-torsional nystagmus is found with no vertical

component if both vertical semicircular canals are equally affected.

4.3.2. Bedside head impulse test (bedside HIT)

Typically, the bedside HIT is pathological with a re-fixation saccade when the head is turned very fast toward the suspected affected side [65]; however, the sensitivity and specificity of the bedside HIT is low [162] and depends on the examiner [102]. Therefore, if the result of the bedside HIT is not clear, a video-HIT (vHIT) and/or caloric testing will be necessary; in addition, if available we recommend that these latter two tests should be performed in all patients to increase the certainty of the diagnosis and to quantify the deficit (see below).

4.3.3. Examination for central ocular motor disorders

There should be no central ocular motor signs that suggest an acute central vestibular syndrome (See: Vascular Vertigo and Dizziness: Diagnostic Criteria Consensus document of the Committee for the Classification of Vestibular Disorders of the Bárány Society [164]).

Skew deviation, i.e., a vertical deviation of eye position of vestibular cause, is examined with the cover and alternating cover test. A small skew SD can be present in some patients with an acute unilateral peripheral vestibular lesion (presumably due to damage of utriculo-ocular afferents) [4, 46, 59, 134]. A study with quantitative measurement of SD showed that a small SD ($< 3^\circ$) is seen in about 20% of patients AUVP [84], so should not be used to exclude an AUVP diagnosis; in ACVS, about 30% had a SD and a prominent SD ($> 3.3^\circ$) pointed toward a central lesion. Therefore, the presence of an observable SD using the cover test (cover-uncover test) and alternating cover test at the bedside contravenes but does not rule out a diagnosis of AUVP.

A direction-changing gaze evoked nystagmus (GEN) indicates a central deficit; in a study of 35 patients with AUVP and 12 patients with stroke, none of the patients with AUVP had GEN and one third of patients with strokes had a spontaneous SN in straight-ahead gaze and a pathological GEN, producing the pattern of a Bruns' nystagmus [105]. Note that an un-sustained endpoint nystagmus can also be found in healthy individuals (for Ref. see [45, 94]).

4.3.4. Examination of hearing and otoscopy

Hearing loss can be screened by using the finger rub test [142] or whispered voice test [126]. The type

of the hearing loss can be suspected by using the Rinne and Weber tests. There should be no evidence for acute hearing loss. This has a double implication for the differential diagnosis, namely in terms of Menière's disease and an infarction of the anterior inferior cerebellar artery. The external ear canal can be visualised by otoscopy. There should also be no evidence for herpes zoster, otitis media, or another pathology.

4.3.5. Measurement of subjective visual vertical (SVV)

There is a monocular and binocular pathological deviation of SVV toward the affected ear reported in patients with an AUPV [19, 41]. A deviation of SVV, however, does not discriminate between a peripheral and a central lesion.

4.3.6. Romberg Test

Postural imbalance on the Romberg test, increased with the eyes closed, typically shows falling toward the slow phase of nystagmus, i.e. the affected side in the acute stage [22].

4.4. Laboratory examinations

Since the bedside HIT is not very reliable in diagnosing a vestibular deficit [162], laboratory tests are often necessary to quantify the function of the VOR and should be used whenever possible. There is a high rate of false-positives and false-negatives when using bedside tests in acute vestibular syndromes [102].

4.4.1. Video-head impulse test (vHIT)

The vHIT [15, 63, 100] measures the function of the angular VOR in the high-frequency range. The testing is performed close to the functionally relevant stimulation range of the semicircular canals, i.e., from 0.1 to 10 Hz. Further, the HIT tests pairs of semicircular canals in both ears rather than isolated semicircular canals in each ear. For example, during a head turn to the right, there is an excitation of the right horizontal and an inhibition of the left horizontal semicircular canal. Since in almost all cases of a peripheral vestibular deficit the horizontal canal is affected, the examination of this canal by the vHIT is clinically sufficient in most patients, except for those with a suspected "inferior vestibular neuritis".

For the calculation of VOR gain, different algorithms are used, e.g. the angular eye velocity is divided by the angular head velocity at 60 milliseconds or the area under the curve of the eye angular

velocity is divided by the area under the curve of head angular velocity. Despite these different approaches to the analysis, the calculated values are practically identical [93, 115, 145]. Depending on the position of the camera in front of the examined eye, there is a right-left difference of the VOR gain, which has not been well explained so far. For instance, if the camera is placed in front of the right eye, the gain to the right is about 5% higher than the gain to the left [145].

The applied normative values/reference range of the HIT are mainly based on studies with a large number of healthy subjects and for different age groups (e.g. [11, 110, 111, 160]). A VOR gain above 0.8 is often classified as normal but there is variability of what is assumed to be normal or pathological [144].

For a significant VOR deficit, the gain should be less than 0.7 and the side-difference greater than 0.3. There should also be saccades during the test. As vHIT measurements are still under development and there is no gold standard to calculate gains, there is a risk of inexact values. Thus, VOR gains estimated using vHIT should be seen as approximations dependent on the equipment and techniques used [158].

The vHIT is also helpful in differentiating between peripheral and central vestibular lesions [31, 101, 106, 120, 133] because a normal vHIT is not compatible with AUPV.

4.4.2. Caloric testing

Caloric testing allows a quantification of the function of a single horizontal semicircular canal in the low, non-physiological, frequency range of about 0.003 Hz by irrigation of the external auditory canal with 30°C cold and 44°C warm water, while induced eye movements are recorded (for references, see [139]). The peak slow phase velocity of the caloric-induced nystagmus is quantified by means of video-oculography. An advantage of this technology is that each horizontal canal can be examined separately, in contrast to video-HIT and rotatory chair testing.

Since there is large interindividual variability of the nystagmus induced by caloric irrigation and less intraindividual variability of the response of the right and the left labyrinths, "Jongkees's formula for vestibular paresis" [74]:
$$\frac{((R30^\circ + R44^\circ) - (L30^\circ + L44^\circ))}{(R30^\circ + R44^\circ + L30^\circ + L44^\circ)} \times 100$$
 should be used to determine its presence. In this formula, for instance, R30° is the peak slow phase velocity during caloric irrigation with 30°C water of the right ear.

Vestibular paresis is usually defined as >25% asymmetry between the two sides [69] or an absolute value for the sum of the mean peak slow phase of the irrigation with warm and cold water <6°/s. It is important to note that in rare “inferior vestibular neuritis”, which affects the posterior semicircular canal only (see below), caloric testing and the vHIT for the horizontal semicircular canal are typically normal.

4.5. Complementary laboratory examinations

4.5.1. Cervical and ocular VEMP

In contrast to vHIT and caloric testing, vestibular evoked myogenic potentials (VEMP) are much less relevant for the diagnosis of AUVP (for references, see [51]). Cervical VEMP (cVEMP) and ocular VEMP (oVEMP) help to differentiate between superior AUVP (the most frequent form) and inferior AUVP and complete AUVP (affecting the superior and the inferior vestibular nerve and thus all semicircular canals and both otolith organs). oVEMP [129], which mainly evaluates the function of the utricle, is typically reduced or absent in superior and complete AUVP [38, 122], since the superior branch of the vestibular nerve innervates the utricle (see below); cVEMP, which mainly evaluates the function of the saccule and is innervated by the inferior vestibular nerve, is reduced or absent in inferior and complete AUVP [32, 37, 108, 116, 122]. Acute isolated utricular or saccular vestibulopathy which is characterized by acute onset of postural imbalance, can also be diagnosed by c/oVEMP.

4.5.2. Imaging

If the patient history and/or the clinical examination provide evidence of an acute central vestibular syndrome (ACVS), immediate imaging is indicated. Computed tomography (CT) can be used to rule out a haemorrhage while CT angiography can diagnose vertebral or basilar artery stenosis. It should be noted that diffusion-weighted MRI (DWI) in small brainstem or cerebellar lesions can be normal within the first 24 hours after symptom onset in up to 16% of patients [140]. Although the sensitivity of imaging in detecting small cerebrovascular lesions can be improved by new diffusion techniques using sagittal and coronal 3 mm slices [48, 123], the rate of false negative DWI supports the relevance of a systematic patient history and the clinical bedside examination of the vestibular and ocular motor systems to look for central signs, because combining history and physical examination permits a differentiation between AUVP

and ACVS with a sensitivity and a specificity of about 90% [36, 75, 133].

Imaging studies can also demonstrate contrast enhancement of the vestibular nerve, in particular the superior vestibular nerve, one to four hours after gadolinium injection [26, 47, 153]. Finally, one study showed that high-resolution MRI examinations ≥ 6 months after symptom onset in patients with a persistent peripheral vestibular deficit revealed atrophy of the vestibular nerve in 5 of the 10 patients examined, in particular of the superior vestibular nerve [53].

4.5.3. Audiogram and otoscopy

If there is evidence from the patient history and/or the clinical examination of an impairment of hearing (see above), otoscopy should be performed to rule out impacted cerumen as a cause of hearing loss. Then an audiogram is indicated in particular since patients with an attack of Menière’s disease and patients with an anterior inferior cerebellar artery (AICA) infarction show an impairment of hearing. Also otoscopy is indicated in every patient with acute vertigo, to look for herpes zoster or otitis media.

4.6. Differential diagnosis

A diagnosis of AUVP is a diagnosis of exclusion. The most important differential diagnoses are (Table 1):

- Acute central vestibular syndrome due to a lesion, most often an infarction, in the brainstem or the cerebellum
- Combined acute central and peripheral lesions, for instance due to an AICA infarction, which can affect the labyrinth, cerebellum and/or brainstem, [88] or Susac syndrome [82]
- Other central vestibular syndromes, e.g. first attack of vestibular migraine
- Other inner ear diseases, e.g. first attack of Menière’s disease or herpes zoster infection, with or without rash (Zoster sine herpette) [18].

4.6.1. Acute central vestibular syndrome (ACVS)

An ACVS, which can mimic AUVP, can occur due to: (a) A lesion in the root entry zone of the vestibular nerve in the lateral medulla (e.g. in multiple sclerosis or small lacunar strokes [151]), causing a fascicular nerve lesion (formerly named pseudoneuritis), (b) A small lacunar infarction of the vestibular nuclei [79] or the dorsolateral pons affecting the cerebellar peduncle [28], (c) A small cerebellar infarction, e.g.,

Table 1

Differential diagnoses for AUVP. Central and peripheral vestibular disorders (in alphabetical order)

Diagnosis	Clinical characteristics
Central vestibular disorders	
Acute central vestibular syndrome	Acute onset and duration often similar to AUVP More often vascular risk factors Often accompanied by central neurological symptoms and signs HINTS-plus important for the differential diagnosis
Vestibular migraine	Acute or subacute onset of often > 24 hrs of symptoms Not always associated with headache Episodes often associated with spontaneous and/or positional nystagmus [155]
Peripheral vestibular disorders	
Cogan's syndrome	Double triad: – Vertigo, bilateral hearing loss/tinnitus, eye pain/ "red eyes" – Bilateral, often asymmetric peripheral vestibular deficits, bilateral hypoacusis, interstitial keratitis Symptoms or deficits often rapidly progress Mostly in young women
Cupulolithiasis of a horizontal semicircular canal	Clinical examination may reveal horizontal pseudo-spontaneous nystagmus, which changes direction during the lean-and-bow test [35] and shows apogeotropic positional nystagmus during positional manoeuvres [154]).
Herpes zoster oticus (Ramsay Hunt syndrome)	Initially burning ear pain and blisters, vertigo, hearing disorders and facial paresis; the symptoms can begin before the skin rash or even without a skin rash (Zoster sine herpette) Can lead to complete AUVP, including skew deviation [4] Often contrast enhancement of the affected cranial nerves on MRI
Isolated acute unilateral utricular or saccular vestibulopathy	Acute onset of postural imbalance which can be diagnosed by ocular and cervical VEMP; repeated testing is recommended to confirm the diagnosis
Labyrinthitis	Associated with ear pain, reduced hearing and/or tinnitus. The course of the disease can be acute, subacute or slowly progressive.
Menière's disease	Can begin with major vestibular and only minor audiological symptoms and is also associated with a nystagmus. The differential diagnosis is difficult and can only be made during the course of the disease (see diagnostic criteria).
Recurrent vestibulopathy	First attack is similar to AUVP, duration often shorter than in AUVP. However, if symptoms re-occur, this differential diagnosis as well as vestibular migraine should be considered; its etiology is still not clear [43].
Vestibular schwannoma	Often a slowly progressive course of the disease and associated with impairment of hearing and/or tinnitus. Often diagnosed in patients who have an asymmetric impairment of hearing by means of contrast-enhanced MRI. Vestibular symptoms may occur only in the later stage of the disease. In rare cases, it may also be associated with attacks of vertigo.
Combined central and peripheral disorders	
AICA infarction	Can affect the labyrinth, cerebellum and/or brainstem (74)
Susac syndrome	Can rarely begin as an acute vestibular syndrome. During the course of the disease, there is a broad spectrum of ophthalmological signs [82].

of the flocculus, which can also cause a pathological HIT [125].

A sensitive and specific instrument to differentiate between AUVP and an ACVS is the combination of the patient history, with particular consideration of the ABCD² score (Age, Blood pressure, Clinical features, Diabetes and Duration; for references, see [40]), and the clinical examination, in particular the HINTS (Head Impulse, Nystagmus, Test of Skew) and the HINTS-plus (Head Impulse, Nystagmus, Test of Skew, finger rub hearing test) protocol [36, 75, 133] to rule out central signs (for references, see [76, 92, 163]). The following aspects of the patient history would support a central origin: Cardiovascular

risk factors, such as arterial hypertension, diabetes, nicotine consumption; age > 60 years; double vision; hemiparesis; hypoesthesia; or hemiataxia.

The following clinical signs (HINTS and HINTS-plus support a central origin [36, 75, 133]: 1. Vertical deviation/skew deviation: If prominent (> 3.3°) this is a sign of a central lesion with a very low sensitivity (found in only 30% of patients with an acute brainstem or cerebellar lesion [21, 84]; a small skew SD can be present in about 20% of patients with an AUVP (presumably due to damage of utriculo-ocular afferents) [4, 46, 59, 134] (see above). 2. Type of spontaneous nystagmus: a spontaneous nystagmus that is not reduced or suppressed by visual fixation

is not a peripheral vestibular spontaneous nystagmus ([107], see for Ref. [66] and above). A pure vertical or a pure torsional nystagmus indicates a central lesion. A pure horizontal nystagmus can indicate a central lesion or can also be found in horizontal canal BPPV as a pseudo-spontaneous nystagmus (see below). 3. A gaze-evoked nystagmus in the opposite direction to the fast phase of the spontaneous nystagmus (e.g., Brun's nystagmus, see above). 4. A normal head-impulse test in acute vestibular syndrome with nystagmus is not compatible with a peripheral deficit. It is important to note that the head-impulse test can also be pathological in central lesions, in particular in a fascicular or nuclear lesion or in floccular lesions [125]. 6. Head-shaking nystagmus: If horizontal head-shaking leads to a change in the direction of the nystagmus or to a vertical nystagmus (cross-coupling) [33, 71], this is compatible with a central lesion; however, a more recent study showed that this is not very specific [161].

Testing of hearing has a two-fold implication: first, for the diagnosis of Menière's disease and other diseases that affect hearing and second, for the diagnosis of an infarction of the anterior inferior cerebellar artery.

Isolated clinical signs, such as skew deviation or a normal HIT are specific, but not very sensitive [36, 75, 119]. The combination of these clinical signs has a sensitivity and specificity of 80–95% [30, 36, 75, 119]. A meta-analysis (5 studies, 617 patients) concluded that the findings also depend on who is performing the examination [124]: Neurologists who frequently used HINTS had a sensitivity of 96.7% and a specificity of 94.8%. Physicians, including neurologists, working in an ENT department had a sensitivity of 83% and a specificity of only 44%. This highlights the importance of combining the patient history, ABCD²-score, and practicing the HINTS.

The vHIT can also contribute to the differential diagnosis: a bilaterally normal, a bilaterally reduced, or a bilaterally increased VOR gain together with a spontaneous nystagmus or crossed vertical refixation saccades during horizontal vHIT are central signs [34].

Cerebellar infarctions. Infarctions in the territory of the posterior inferior cerebellar artery (PICA) and the anterior inferior cerebellar artery (AICA) can manifest as ACVS. This is particularly true for PICA infarctions [44, 70, 103, 104], which can lead to isolated infarctions of the nodulus [113]. Cerebellar infarctions can also cause an incomplete ocular

tilt reaction (OTR) [114], in particular if the dentate nucleus [12], flocculus [125], nodulus [91], tonsil, uvula, or middle cerebellar peduncle [12] is affected.

Infarctions in the AICA territory, which supplies the cerebellum, brainstem, and inner ear, can manifest as ACVS with strong lateropulsion and hearing impairment in combination with central symptoms and signs [89, 90]. It should be noted that they are often associated with acute severe unilateral hearing loss due to an infarction in the territory of the labyrinthine artery, which supplies the cochlea, the semicircular canals, and the otolith organs.

Vestibular migraine. The leading symptom of vestibular migraine based on current diagnostic criteria [96] is the acute onset of vertigo that can last for up to 72 h and can be associated with spontaneous peripheral and central vestibular as well as positional nystagmus [156]. Therefore, the differential diagnosis can be difficult, in particular in a first attack of vestibular migraine and in the elderly in whom attacks of vestibular migraine occur more often without headache. Helpful criteria are a history of migraine, typical migrainous symptoms with the attack of vertigo and the application of the HINTS criteria (see above).

4.6.2. Other peripheral vestibular disorders

There are many other peripheral vestibular disorders that can lead to similar symptoms and signs as AUVP (Table 1). Important differential diagnoses are Menière's disease, which can begin with major vestibular and minor audiological symptoms and is also associated with spontaneous nystagmus. The short duration of the attacks and the quick recovery is usually helpful for establishing a diagnosis.

Cupulolithiasis of the horizontal semicircular canal also leads to spinning vertigo and horizontal nystagmus. Besides positional nystagmus, pseudo-spontaneous nystagmus may be observed in the upright head position beating typically to the affected ear (pseudo-spontaneous nystagmus is a form of positional nystagmus that occurs with the head in the upright position, mimicking spontaneous nystagmus). In contrast to spontaneous nystagmus, pseudo-spontaneous nystagmus is strongly influenced by head position and ceases with the head tilted about 30° forward [9]. Furthermore, there is a change in the direction of the horizontal pseudospontaneous nystagmus during the lean-and-bow test [35]. Symptoms and signs in horizontal semicircular canal variants of BPPV occur in attacks that depend on head position with a change in the direction of the

horizontal nystagmus during the lean-and-bow test and during positioning manoeuvres.

Isolated acute unilateral utricular or saccular vestibulopathy is another differential diagnosis, which is characterized by acute onset of postural imbalance, and can be diagnosed by ocular and cervical VEMPs, respectively.

Inflammatory inner ear diseases that are associated with an AUVP include herpes zoster oticus and labyrinthitis. Herpes zoster oticus is usually associated with a skin rash with blisters, but can also manifest before the rash or rarely as *Zoster sine herpette* [18]. It often presents with lesions of other cranial nerves, e.g., the facial nerve, and, like labyrinthitis, it may be associated with ear pain.

Table 1 summarizes the most relevant central and peripheral vestibular differential diagnoses and their clinical features.

4.7. Pathophysiology and pathological anatomy

4.7.1. Static deficits, e.g., spinning vertigo, spontaneous peripheral vestibular nystagmus

Normal vestibular end organs generate an equal resting firing frequency of the axons, which is the same on both sides. This continuous excitation (resting discharge rate in monkey ≈ 100 Hz [58] of 18,000 vestibular afferents for each labyrinth [16], i.e. 1.8 million action potentials per second) is transmitted to the vestibular nuclei via vestibular nerves. Pathological processes affecting an end organ or vestibular nerve alter its firing frequency, thereby creating a vestibular tone imbalance. This causes spontaneous nystagmus with the slow phase (which is the pathological component of the nystagmus) of the eye movement toward the impaired labyrinth. This imbalance is also the cause of other manifestations on different levels, i.e. perceptual (spinning vertigo, ipsilateral displacement of the subjective vertical (for Ref. see [41]), ocular motor (ipsilateral ocular torsion in addition to spontaneous nystagmus), postural (ipsilateral gait deviation or falling), and vegetative (nausea and vomiting). The static deficits diminish over weeks (for Ref see [85]).

4.7.2. Dynamic deficits

Dynamic deficits of the VOR can be demonstrated by the HIT, vHIT, caloric irrigation and rotational testing. If the peripheral vestibular deficit persists, the dynamic deficits do not recover completely because they cannot be entirely compensated.

4.7.3. Superior vestibular nerve, inferior vestibular nerve or total vestibular nerve lesion

The 3-D features of the spontaneous nystagmus and the dynamic deficit of the VOR of the horizontal, anterior, and posterior semicircular canals were measured in patients with “vestibular neuritis” by means of the scleral coil technique and analysed by a vector analysis in 1996 [50]. These measurements supported the earlier view [23] that AUVP is – in most cases – a partial rather than a total unilateral vestibular lesion (see below) most often affecting the superior division of the vestibular nerve only (innervating the horizontal and anterior semicircular canals, the macula of the utricle, and the antero-superior part of the saccule), which has its own path and ganglion [99, 135], whereas the inferior vestibular nerve (innervating the posterior semicircular canal and the postero-inferior part of the macula of the saccule) is most often spared, leading to “superior vestibular neuritis”. These findings were further supported by (a) measurements of the function of individual semicircular canals with the head-impulse test and the scleral coil technique [10, 20, 49] and (b) VEMP studies in which the cVEMP was most often normal but the oVEMP was reduced or absent [32, 37, 108, 116, 122]. The sparing of the inferior vestibular nerve has two implications: first, with respect to clinical findings, because it explains why patients with AUVP can suffer from “post-infectious” benign paroxysmal positional vertigo of the posterior canal; second, with respect to the pathophysiology and etiology because a theory on that has to explain this fact. Reasons for this include the longer and smaller bony canal through which the superior vestibular nerve runs [55], as well as the double innervation of the posterior canal with two nerves: the posterior ampullary and the accessory posterior ampullary nerve [5, 7, 25, 112].

Using 3-D eye movement recordings and cVEMP/oVEMP measurements, the first cases of rare “inferior vestibular neuritis” were described in 1996 and 1997 [20, 116]. The direction of the spontaneous nystagmus corresponded to the plane of the posterior semicircular canal, i.e. it was contraversively torsional (i.e., away from the affected ear) with a downward component (the opposite direction to that which is found in posterior canal BPPV) [80]. It is often misdiagnosed as “central”.

If the superior and the inferior vestibular nerve (“total vestibular neuritis”) are affected, horizontal nystagmus with a torsional component is found with no vertical component because both vertical canals

are affected. Such a nystagmus can also occur in herpes zoster oticus, which can lead to a complete OTR with a skew deviation [4].

4.8. Etiology

A viral etiology of AUVP is – in analogy to Bell’s palsy and some types of acute hearing loss – likely but so far not proven [13, 14, 54, 86, 117, 137]. There are several lines of evidence for such an etiology: a) a post-mortem study showed inflammatory degeneration of the vestibular nerve [136], b) demonstration of HSV-1, latency-associated transcripts in vestibular ganglia, and activated CD8 + T cells [3, 5–8, 68, 150]. c) two genome-wide association studies, which showed single nucleotide variants in the host factor for HSV-1 replication [130] and a high-risk allele for herpes labialis severity [131].

It is assumed that as a result of intercurrent factors, which impair the immune system, HSV-1 replicates and induces an inflammation and oedema and, thereby, secondary cell damage of the vestibular ganglion cells and axons in the bony canals, which may also explain the therapeutic effect of steroids in the very acute phase [77, 149].

The canal of the superior vestibular nerve is longer and has more spiculae than the canal of the inferior vestibular nerve [55, 57], which innervates the posterior semicircular canal. Also, the posterior semicircular canal is innervated by an additional anastomosis [7, 25]. These and other factors including redundancy in innervation of the saccule and posterior semicircular canal and anastomoses between the facial nerve and the superior vestibular nerve through which reactivated herpes virus may spread may explain why the posterior semicircular canal is often but not always spared in AUVP [5, 25].

However, despite these findings, an inflammatory or even viral etiology has not so far been proven and the same symptoms and signs can also be caused by an infarction of the anterior/superior vestibular artery which is more vulnerable than the inferior vestibular artery [24]. The term “acute unilateral vestibulopathy” is preferred, though the term “vestibular neuritis” can be used synonymously.

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