Original Article

Bilateral Vestibular Hypofunction in a Tertiary Dizziness Center: Occurrence and Etiology

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BACKGROUND: The primary goal of this study was to determine the occurrence of bilateral vestibular hypofunction in a specialized dizziness clinic and to assess the etiology in patients diagnosed with bilateral vestibular hypofunction. Secondary goal was to find out if the diagnosis was already made before the patient was seen at our clinic.

METHODS: A retrospective cohort study, including patients who visited our specialized dizziness center between January 1, 2008, and December 31, 2018, fulfilling the criteria for bilateral vestibular hypofunction according to the Classification Committee of the Bárány Society (2017). Data were collected regarding symptoms, causes, and vestibular function.

RESULTS: In total, 126 patients met our initial inclusion criteria, of which 103 patients met the Classification Committee of the Bárány Society criteria for bilateral vestibular hypofunction, so patients with bilateral vestibular hypofunction comprised 0.9% of the total population seen at our clinic. Mean age was 65.2 years and 49.5% were female. In only 29.1% of patients, the diagnosis was already made elsewhere. A definite cause was identified in 39.8%, the most common cause being ototoxicity.

CONCLUSION: About 1% of the patients visiting our dizziness clinic has bilateral vestibular hypofunction. In our patient population, ototoxicity was the most common cause of bilateral vestibular hypofunction, and in more than 40%, the cause remains unknown. In the majority of the cases, the diagnosis of bilateral vestibular hypofunction was first made at our clinic and not by the referring general practitioner or specialist. When using the Classification Committee of the Bárány Society criteria for bilateral vestibular hypofunction and presbyvestibulopathy, some patients with bilateral vestibular weakness and complaints cannot be categorized in either group.

KEYWORDS: Bilateral vestibulopathy, etiology, vestibular function tests

INTRODUCTION

Bilateral vestibular hypofunction (BVH) is a clinical condition defined by an absent or impaired function of the vestibular organs, the 8 cranial nerve, or a combination of both.1 The clinical picture is characterized by oscillopsia—the experience that the environment is moving when the head is moving—and imbalance during motion. The imbalance is worse in poorly illuminated environments or when walking on uneven, spongy ground.1-4 Bilateral vestibular hypofunction may also present with visual vertigo, cognitive deficits, impaired spatial orientation, and/or neurological, auditory, and/or autonomic symptoms.3-5 The symptoms can be disabling, that is 41% of the patients perceive their handicap—measured with the Dizziness Handicap Inventory (DHI)—as moderate and 44% as severe.6 Lucieter et al7 found a mean DHI total score of 56.0 in BVH patients, indicating a moderate handicap. In the literature, prevalence rates for BVH vary from 28 to 81 per 100 000 people.6-9 Hain10 states that 1% of all the patients with dizziness visiting his clinic is diagnosed with BVH. Occurrence or prevalence rates of BVH in the Netherlands are currently lacking.

Several studies have shown that in 49%-80% of patients with BVH, a definite or probable cause can be identified, the most common causes being ototoxicity, bilateral Meniere’s disease (MD), and meningitis (Table 1). Unfortunately, in 20%-51% of the patients,
the cause remains unclear.2,5,12 Multiple studies have focused on this problem, and migraine and/or autoimmunity seem to play a role in the etiology.12-15

Due to the heterogeneous symptomatology of BVH and unfamiliarity with the condition among general practitioners (GPs), BVH is sometimes overlooked which may result in misdiagnosis or a diagnostic delay.1,3,16 For years, it was the norm to diagnose BVH when the sum of the peak slow phase velocity (SPV) of all 4 irrigations was below 20°/s as measured by caloric testing.4,17-20 However, no formal diagnostic criteria existed until 2017, when the Classification Committee of the Bárány Society (CCBS) published diagnostic criteria for BVH (Table 2).21 In addition, in 2019, the CCBS published diagnostic criteria for presbyvestibulopathy (PVP). Presbyvestibulopathy is defined as bilateral vestibular function loss due to aging. It presents with the same symptoms as BVH, but the criteria differ regarding age and outcomes of diagnostic testing (Table 2).22 Therefore, PVP and BVH are considered as 2 separate disorders.

The primary goal of this study was to determine the occurrence of BVH in a specialized dizziness clinic and to assess the etiology in patients diagnosed with BVH. Secondary goal was to find out if the diagnosis was already made before the patient was seen at our clinic.

**MATERIALS AND METHODS**

**Ethics**
The study was conducted in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments.

### Table 1. Etiology of Bilateral Vestibular Hypofunction1,5,12,16

| Categories          | Causes                                                                 |
|---------------------|------------------------------------------------------------------------|
| Idiopathic          | Aminoglycoside antibiotics, some chemotherapeutic agents, furosemide, aspirin, alcohol, vitamin-B12 deficiency, folate deficiency, hypothyroidism, styrene poisoning, combination of NSAID + penicillin |
| Toxic               | Meningitis/encephalitis/cerebellitis, Borrelia, bilateral vestibular neuritis, Lues, Behçet, Herpes simplex virus |
| Infectious          | Sarcoidosis, Cogan, Susac, Sjögren’s, colitis, celiac disease, polyarteritis nodosa, antiphospholipid syndrome, other systemic diseases |
| Autoimmune          | Superficial siderosis, CANSAS, multiple system atrophy, polyneuropathy, episodic ataxia, SCA3, SCA6, hereditary sensory and autonomic neuropathy type IV, other ataxias |
| Neurodegenerative   | Neurofibromatosis type 2, bilateral vestibular Schwannoma, lymphatic metastasis, other malignant tumors |
| Genetic             | DFNA6, DFNA11, DFNA15, DFNB4, mutations on the 5q, 6q, 11q, or 22q chromosome and Muckle–Wells syndrome |
| Vascular            | Vertebrabasilar dolichoectasia, supra- or infratentorial abnormality |
| Neoplastic          | Lateral (e.g., bilateral cochlear implant), head trauma |
| Trauma              | Otosclerosis, cholesteatoma, or bilateral labyrinthitis |
| Other ear pathology | CHARGE, Turner, Usher, Alport syndrome, enlarged vestibular aqueduct syndrome |
| Congenital          | Vestibular atelectasis, presbyvestibulopathy, auditory neuropathy spectrum disorders |
| Other               | BVH, bilateral vestibular hypofunction; CANSAS, cerebellar ataxia with neuropathy and bilateral vestibular areflexia syndrome; SCA, spinocerebellar ataxia; CHARGE, colomba, heart defects, atresia of the choanae, retardation of growth and development, genital and urinary abnormalities, ear abnormalities and/or hearing loss DFNA, deafness autosomal dominant inherited hearing loss; DFNB, deafness autosomal recessive hearing loss; NSAID, non-steroidal anti-inflammatory drug. |

### Table 2. Diagnostic Criteria21,22

| BVH                                           | PVP                                           |
|---------------------|------------------------------------------------|
| **A.** Chronic vestibular syndrome with the following symptoms: | **A.** Chronic vestibular syndrome (at least 3 months duration) with at least 2 of the following symptoms: |
| 1. Unsteadiness when walking or standing plus at least 2 or 3 | 1. Postural imbalance or unsteadiness |
| 2. Movement-induced blurred vision or oscillopsia during walking or quick head/body movements and/or | 2. Gait disturbance |
| 3. Worsening of unsteadiness in darkness and/or on uneven ground | 3. Chronic dizziness |
| | 4. Recurrent falls |
| **B.** No symptoms while sitting or laying down under static conditions | **B.** Age ≥ 60 years |
| **C.** Bilaterally reduced or absent VOR function documented by: | **C.** Mild, bilateral peripheral vestibular hypofunction documented by at least 1 of the following: |
| 1. Bilateral pathological horizontal angular VOR gain < 0.6, measured by the video head impulse test or sclera-coil technique and/or | 1. VOR gain measured by video-HIT between 0.6 and 0.8 bilaterally |
| 2. Reduced caloric response (sum of bithermal max. peak slow-phase velocity) on both side <6°/sec) and/or | 2. VOR gain between 0.1 and 0.3 upon sinusoidal stimulation on a rotatory chair (0.1 Hz, Vmax = 50-60°/sec) |
| 3. Reduced horizontal angular VOR gain <0.1 upon sinusoidal stimulation on a rotatory chair and a phase lead > 68° (time constant < 5 seconds) | 3. Reduced caloric response (sum of bithermal maximum peak SPV on each side between 6 and 25°/sec) |
| **D.** Not better accounted for by another disease | **D.** Not better accounted for by another disease |

HIT, Head Impuls Test; VOR, vestibular–ocular reflex; SPV, slow-phase velocity.
and 4 points, total score ranges from 0 to 100. Scores between 0 and 30 are considered to be mild, between 31 and 60 to be moderate, and between 61 and 100 to be severe.26

Patients were included in this study if they visited our clinic between January 1, 2008, and December 31, 2018, and met the criteria for BVH applicable at that time, that is (1) experienced imbalance during movement and/or oscillopsia and (2) had a reduced caloric response—summated mean peak SPV of 20°/s—and/or (3) a reduced gain during video head impulse testing (vHIT)—average gain of ≤0.6.

Patients were excluded if the outcomes of the vHIT or caloric testing were not available and/or if they had a unilateral vestibulopathy which was defined as a vestibular preponderance (VP) of 22% or higher.

After inclusion, we applied the CCBS criteria for BVH—as shown in Table 2—to our population and analyzed only those patients who met the new criteria for BVH.

First, we collected the date on which the diagnosis of BVH was established in our clinic. If patients were seen multiple times in our clinic, the date of the first consultation at which the diagnosis of BVH was made was noted. Second, we determined from the information in the patient file if BVH was already diagnosed—based on vestibular function tests—elsewhere or suspected before visiting our clinic. Third, we determined if the referral was a second opinion or a primary referral from a GP.

Imbalance symptoms were defined as feeling dizzy, feeling light-headed, and experiencing imbalance during motion. Oscillopsia was defined as blurry vision during motion and the experience of seeing multiple images during motion.

When the patient received one or more sessions of vestibular rehabilitation at our hospital, then in the context of usual care, the impact of dizziness on daily life was measured by the Dutch version of the DHI. Total DHI score pre-treatment was collected and categorized.23,24 The DHI is a 25-item questionnaire to assess the impact of the DHI. Total DHI score pre-treatment was collected and categorized.23,24 The DHI is a 25-item questionnaire to assess the impact of the DHI. Total DHI score pre-treatment was collected and categorized.23,24 The DHI is a 25-item questionnaire to assess the impact of the DHI. Total DHI score pre-treatment was collected and categorized.23,24 The DHI is a 25-item questionnaire to assess the impact of the DHI. Total DHI score pre-treatment was collected and categorized.23,24 The DHI is a 25-item questionnaire to assess the impact of the DHI. Total DHI score pre-treatment was collected and categorized.23,24 The DHI is a 25-item questionnaire to assess the impact of the DHI. Total DHI score pre-treatment was collected and categorized.23,24 The DHI is a 25-item questionnaire to assess the impact of the DHI. Total DHI score pre-treatment was collected and categorized.23,24 The DHI is a 25-item questionnaire to assess the impact of the DHI. Total DHI score pre-treatment was collected and categorized.23,24 The DHI is a 25-item questionnaire to assess the impact of the DHI. Total DHI score pre-treatment was collected and categorized.23,24 The DHI is a 25-item questionnaire to assess the impact of the DHI. Total DHI score pre-treatment was collected and categorized.23,24 The DHI is a 25-item questionnaire to assess the impact of the DHI. Total DHI score pre-treatment was collected and categorized.23,24

Table 3 shows characteristics of our patient population. The mean age of the BVH patients was 65.2 ± 14.5 years (range, 25-89). Forty-six patients (44.7%) were diagnosed with BVH between the age of 51 and 70. Half of the BVH patients were female (49.5%). The mean average gain of the vHIT was 0.4 ± 0.2, and the mean maximum SPV was 7.4 ± 8.6. A total of 37 patients (35.9%) completed the DHI, of whom 16 rated their handicap due to dizziness to be moderate (43.3%).

In 43 of the 103 (41.7%) BVH patients, a cause could not be identified. A definite cause was identified in 41 (39.8%) of the patients, and a probable cause in 19 (18.5%) of the patients.

Idiopathic BVH is the largest category followed by ototoxicity (Figure 2). Twenty out of 29 patients in the toxic medication group had a history of gentamicin administration (19.4%). In 4 patients (3.9%), vancomycin, tobramycin, or other chemotherapeutic agents (e.g., cisplatin or carboplatin) were identified as the cause. A suspicion of toxic medication was present in 5 (4.9%) patients. After toxic medication (28.2%), the most frequent causes were meningitis (n = 5; 4.9%) and cerebellar ataxia, neuropathy and vestibular areflexia syndrome (CANVAS) (n = 5; 4.9%) (Figure 3).
In 29.1% (n = 30), the diagnosis of BVH was confirmed in our clinic. A total of 22 patients were referred for a second opinion, of whom 7 patients had a suspicion of BVH. The remaining group of 39 patients was referred by their GP for diagnostic testing and treatment. In 11.7%, a referral letter was absent.

A total of 126 patients met our initial inclusion criteria, of whom 103 patients met the 2017 CCBS criteria for BVH. This comes down to a “misdiagnosis” of 23 patients. In hindsight, 17 of these patients (13.5%) could be diagnosed with PVP according to the 2019 diagnostic criteria for PVP. This leads up to 6 patients (4.8%) who neither met the diagnostic criteria for BVH nor for PVP (“Not-BVH-or-PVP”—NBP—group) (Figure 1). Patient characteristics, outcomes on vestibular functions tests, and DHI in the PVP and NBP subgroups are also shown in Table 3.

**DISCUSSION**

The aim of this study was to determine the occurrence and etiology of BVH in our specialized dizziness clinic. Secondary goal was to find out if the diagnosis was already established elsewhere. In summary, BVH was present in 103 patients, which comprises 0.9% of the total patient population of our dizziness clinic. The most common causes were idiopathic BVH, followed by ototoxic medication, meningitis, and CANVAS. In only 29.1%, BVH was diagnosed before the referral to our specialized dizziness clinic. Despite meeting our initial inclusion criteria, 103 patients met the 2017 CCBS criteria for BVH. This comes down to a “misdiagnosis” of 23 patients. In hindsight, 17 of these patients (13.5%) could be diagnosed with PVP according to the 2019 diagnostic criteria for PVP. This leads up to 6 patients (4.8%) who neither met the diagnostic criteria for BVH nor for PVP (“Not-BVH-or-PVP”—NBP—group) (Figure 1). Patient characteristics, outcomes on vestibular functions tests, and DHI in the PVP and NBP subgroups are also shown in Table 3.

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**Figure 1.** Flow diagram of inclusion of patients with bilateral vestibular hypofunction and distribution of diagnoses. n, number of patients; BVH, bilateral vestibular hypofunction; PVP, presbyvestibulopathy; NBP, not-BVH-or-PVP.

**Figure 2.** Distribution of etiology of bilateral vestibular hypofunction in main categories. BVH, bilateral vestibular hypofunction. Main categories are shown in Table 1.

**Table 3. Patients Characteristics**

|                  | Initial Group (n = 126) | BVH (n = 103) | PVP (n = 17) | NBP (n = 6) |
|------------------|-------------------------|---------------|--------------|-------------|
| Sex, n (%)       |                         |               |              |             |
| Female           | 65 (51.6)               | 51 (49.5)     | 9 (52.9)     | 5 (83.3)    |
| Male             | 61 (48.4)               | 52 (50.5)     | 8 (47.1)     | 1 (16.7)    |
| Age              |                         |               |              |             |
| Mean ± SD        | 64.7 ± 15.9             | 65.2 ± 14.5   | 73.0 ± 9.0   | 32.1 ± 16.1 |
| Range            | 20-90                   | 25-89         | 60-90        | 20-63       |
| VmaxCO*          |                         |               |              |             |
| n                | 103                     | 80            | 17           | 6           |
| Mean ± SD        | 9.3 ± 8.5               | 7.4 ± 8.6     | 16.8 ± 3.1   | 14.2 ± 1.9  |
| Average gain vHIT** |                   |               |              |             |
| n                | 90                      | 79            | 6            | 5           |
| Mean ± SD        | 0.4 ± 0.2               | 0.4 ± 0.2     | 0.7 ± 0.1    | 0.8 ± 0.1   |
| DHI***           |                         |               |              |             |
| Mean ± SD        | 53.0 ± 21.5             | 53.5 ± 22.0   | 54.8 ± 18.1  | 26.0        |
| DHI severity, n |                         |               |              |             |
| Mild             | 9                       | 7             | 1            | 1           |
| Moderate         | 18                      | 16            | 2            |             |
| Severe           | 16                      | 14            | 2            |             |

BVH, bilateral vestibular hypofunction; DHI, Dizziness Handicap Inventory; NBP, no BVH or PVP; PVP, presbyvestibulopathy; SD, standard deviation; SPV, slow-phase velocity

*The summated peak SPV of all four irrigations measured by caloric testing
**The average score of the gain on both sides measured by vHIT
***Number of patients for whom a DHI score is available in the initial group n = 42, BVH group n = 37, PVP group n = 5, NBP group n = 1.
criteria, 6 patients with complaints of imbalance and/or oscillopsia did not meet the newer CCBS criteria for BVH or PVP (the NBP group).

Hain found that in his medical practice, which specializes in dizziness, about 1.0% of all dizziness is due to BVH. This is almost identical to the percentage of BVH patients in our clinic (0.9%). Occurrence rates of BVH specifically for the Netherlands are lacking. The study of Lucieer et al conducted in the Netherlands is comparable to our study. They included 154 BVH patients in 2 years; however, it is unclear what the total population of patients was at their clinic; therefore, the occurrence rate could not be calculated.

The etiology of BVH was classified as idiopathic in 41.7% of the patients, a definite cause was identified in 39.8%, and a probable cause in 18.5%. These findings are largely in line with the findings in other studies. The most common identified causes were ototoxic medication, meningitis, and CANVAS. Rinne et al found a similar distribution; however, in their study, CANVAS was more frequent than meningitis. In the studies conducted by Lucieer et al and Zingler et al, bilateral MD had a more prominent role in the list of most common causes. In our study, bilateral MD was identified in only 2 patients. Furthermore, compared to Lucieer et al, genetic causes were less common in our study. Because our hospital does not have the opportunity to do genetic analysis, patients with a suspicion of the DFNA-9 mutation were referred to another (university) hospital, mostly by their GP. Feedbacks regarding these outcomes were not available and therefore classified as a probable cause. In 4 cases, we indicated aging as the cause of BVH.

Approximately one-third (35.9%) of the BVH patients had a certain diagnosis or suspicion of BVH when referred to our clinic. In two-thirds of the patients, a diagnosis of BVH was lacking, which illustrates the difficulty of diagnosing BVH and the need to refer to a tertiary center. As far as we know, so far no research has been done to assess if the diagnosis was already made before the patient was seen at a specialized dizziness clinic. Unfortunately, we did not collect data to specify the delay in diagnosis.

Our study shows the clinical consequences of the recently published diagnostic criteria for BVH and PVP. In total, 23 patients who were initially classified as having BVH did not meet the CCBS criteria for BVH. Seventeen patients could be classified as having PVP instead of BVH, and 6 patients (the NBP group) met the criteria for neither BVH nor PVP. The NBP group is a small group comprising relatively young, mostly female patients with weakness at caloric testing, and an average gain of 0.8 at vHIT. Dizziness Handicap Inventory scores in the PVP group are comparable to BVH group (54.8 vs. 53.5 points). This level of handicap is comparable to other studies where the average DHI score of patients with BVH varies from 46.9 to 62.0 points. Inner hair cells of the vestibular organ do not regenerate; therefore, spontaneous recovery is unlikely, and half of the BVH subtypes have a progressive nature, which means that the symptoms in the PVP and NBP group can progressively worsen over time. This highlights the need of an adequate and early diagnosis and thereafter adequate information about the condition and its course and possible treatment options in all 3 groups.

In the CCBS consensus document with diagnostic criteria for BVH, a statement is made about the stringency of the criteria: the new diagnostic criteria should be considered as "profound" BVH and less dramatic outcomes of vestibular function tests as "severe" BVH. In addition to this, the authors state that the summated maximum SPV of all 4 irrigations of 20°/sec or less is sensitive but not specific enough because of anatomical differences. As a result, CCBS decided to use stricter diagnostic criteria regarding caloric testing. Starkov et al (2021) published an update on diagnosing vestibular hypofunction. They stated that there is still no worldwide consensus with respect to a standardized testing procedure and normative values for the vHIT and caloric testing. Values of both the vHIT and caloric testing depend to a great extent on the training and experience of
technicians, as well as the equipment used. Laboratories are therefore advised to determine specific normative values for their own setting; however, these values are often lacking. Therefore, in our opinion, the current CCBS criteria for BVH and PVP should be applied with this footnote in mind.

The strength of the current study is the long study period of 11 years and the rather large cohort size. Due to changing diagnostic criteria, we were able to show the clinical consequences of the new diagnostic criteria for BVH and PVP. The retrospective nature of the study is one of the limitations that may have influenced the results. The etiology was not determined in the same way in all the patients. For example, laboratory testing of blood samples was not done on every patient. As a consequence, the size of our idiopathic group could be overestimated. We encountered missing and incomplete data. Unfortunately, only a minority of the BVH patients had completed the DHI, and some patients had to be excluded due to missing results regarding vestibular function testing.

CONCLUSION
About 1% of the patients visiting our dizziness clinic has BVH. In our patient population, ototoxicity was the most common cause of BVH, and in more than 40%, the cause remains unknown. In the majority of the cases, the diagnosis of BVH was first made at our clinic and not by the referring GP or specialist. When using the CCBS criteria for BVH and PVP, some patients with bilateral vestibular weakness and complaints cannot be categorized in either group.

Ethics Committee Approval: Ethical committee approval was received from Gelre Hospitals Institutional Review Board (no: 2020-14).

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