
DISORDERS

NO FUNCTION

When the vestibular system doesn't function, you experience a complete loss of balance.

ARTICLE

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Bilateral Vestibular Hypofunction

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Reduction or loss of vestibular function bilaterally results in difficulty maintaining balance, especially when walking in the dark or on uneven surfaces, and in a decrease in the patient's ability to see clearly during head movements. In addition, patients with bilateral vestibular hypofunction or loss (BVH or BVL) also complain of intense feelings of being off-balance and of strange but disturbing sensations in their heads with head movement. Because of these problems, patients with BVH may restrict their activities and can become socially isolated.

Vestibular rehabilitation can improve postural stability, decrease the sense of disequilibrium, and improve visual acuity during head movements enabling people with BVH to resume a more normal life.¹⁻⁵ Unfortunately, most patients have residual functional problems and subjective complaints.⁵ The exercises used for patients with BVL are aimed at fostering the substitution of alternative strategies to compensate for the lost vestibular function and at improving any remaining vestibular function.

ETIOLOGY

Bilateral vestibular hypofunction and loss can occur as secondary to a number of different problems including ototoxicity, meningitis, sequential vestibular neuritis, progressive disorders, autoimmune disorders, chronic inflammatory peripheral polyneuropathy, congenital loss, and neurofibromatosis. In most cases, however, BVH is considered to be "idiopathic" because the underlying cause cannot be identified. The incidence of the various forms of BVH is also not clear, because it varies depending on the type of clinical practice examined.

PRIMARY COMPLAINTS

Balance and Risk for Falling

Patients with bilateral vestibular loss are primarily concerned with their balance and gait problems. During the acute stage, they may feel off



balance even when lying or sitting down. More typically, however, their balance problems become obvious only when they are standing or walking. Thus, patients who are bedridden, such as those who develop BVH after receiving a vestibulo-toxic medication such as gentamicin, often do not know they have a balance problem until they get out of bed. Typically, these patients have been treated with the ototoxic medication because of a serious infection. They are often debilitated, and their balance problems are initially attributed to weakness. Other patients, such as those with a progressive loss, may not notice their gradually worsening balance until it reaches a critical point and they start falling.



Even with full compensation, balance problems will persist. Although the other sensory and motor systems do help compensate for the vestibular loss, these systems cannot substitute completely for the loss of vestibular function. Normal postural stability while walking requires the combined use of at least two of three sensory cues (visual, vestibular, somatosensory). Patients who have no vestibular function, therefore, will have difficulty when either visual or somatosensory cues are also significantly decreased (e.g., walking in the dark). Although balance may be poor, it is not known what the actual frequency of falling is for patients with BVL. One study reported that 70% of patients with BVH under the age of 65 reported falls related to their bilateral hypofunction and 58% of those 65 to 74 years of age reported falls related to their BVH.⁶ In both cases the incidence of falls exceeded the fall rate of healthy individuals of the same age. Interestingly, patients age 75 years and older had a lower fall rate than do healthy individuals; this was attributed to their use of assistive devices and limited activity level. Most patients are able to prevent falls even though they may side-step or

stagger occasionally. However, even after a course of vestibular and balance re-habilitation, 20% to 30% of patients with BVH are still at risk for falling when they are discharged.^{3,7}

Oscillopsia

Another problem for patients with bilateral vestibular loss is the visual blurring that occurs during head movements. Almost 70% of all patients with BVH complain of oscillopsia even after a course of gaze stabilization exercises.⁷ Greater intensity of oscillopsia occurs in patients with absence of both inferior and superior vestibular nerve function. In addition to the subjective complaint, patients also have a documented decrement in visual acuity during head movements. Interestingly, several studies have demonstrated that the subjective complaints of oscillopsia does not correlate with the actual decrement in visual acuity during head movement.^{5,8,9}

Initially, loss of vestibular function results in a decrement in visual acuity even when the patient is stationary, if the head is not supported¹⁰, and even following the best compensation some patients say that objects that are far away appear to be jumping or bouncing. This visual blurring, or oscillopsia, increases with irregular or unpredictable head movements such as would occur while walking. As a result, patients may not be able to read street signs or identify people's faces as they walk, or they may have difficulty seeing clearly while in a moving car. Severe oscillopsia will also impact postural stability because decreased visual acuity will affect the person's ability to use visual cues for stability.¹¹

DIAGNOSTIC CRITERIA

In 2017 the Bárány Society, an expert panel committed to vestibular research, formulated diagnostic criteria for bilateral vestibulopathy. Details about the process have been refined in the following criteria¹¹:

- Chronic vestibular syndrome with the following symptoms
 - Unsteadiness when walking or standing plus at least one of #2 and #3
 - Movement-induced blurred vision or oscillopsia during walking or quick head/body movements and/or
 - Worsening of unsteadiness in darkness and/or uneven ground
- No symptoms while sitting or lying down



under static conditions

- Vestibular testing reveals bilaterally reduced or absent angular vestibulocular reflex (VOR) function documented by
 - Video head impulse tests (vHIT) or scleral-coil technique and/or
 - Caloric tests and/or
 - Rotational chair tests
- Not better accounted for by another disease

ASSESSMENT

Physical therapy assessment of patients with BVL must address subjective complaints, postural instability and oscillopsia, the patient's overall physical condition, and their ability to perform activities of daily living (ADLs). This assessment must also identify other factors that might affect recovery, especially visual and somatosensory deficits.

Vestibular Function

One important consideration in designing a treatment program is whether there is any remaining vestibular function. Vestibular function can be documented using tests such as the rotational chair and caloric tests. The presence of remaining vestibular function can be used as a guide in predicting the final level of recovery for patients.^{2,3,7} Patients with incomplete bilateral vestibular loss are often able to return to activities such as driving at night and to some sports. Patients with severe bilateral loss may not be able to drive at night, and some patients will not be able to drive at all because of the gaze instability. Activities such as sports and dancing may be limited because of vision and balance problems.

In certain cases, vestibular function tests can also be used to follow the course of the vestibular loss and of any recovery of vestibular function that might occur.^{12,13} Certain aminoglycoside antibiotics are selectively taken up by vestibular hair cells and result in a gradual loss of vestibular function. Typically, there is continued loss of vestibular function even after the medication is stopped. Some improvement in vestibular function may occur in hair cells that were affected by the ototoxic drug but did not die. Potentially, an increase in gain may also occur with the use of vestibular adaptation exercises. This has been demonstrated in patients with unilateral vestibular loss but not in patients with BVL.^{14,15}

MECHANISMS OF RECOVERY

The mechanisms used to stabilize gaze in the absence of vestibular inputs have been well studied. The mechanisms involved in maintaining postural stability are still somewhat less well understood, although research is being done in that area.

Visual and somatosensory cues will not substitute fully for the lost vestibular contribution to postural stability.¹⁶⁻¹⁸

Compensatory Strategies

Patients can be taught, and often develop on their own, strategies to use when in situations in which their balance will be stressed. For example, a patient may learn to turn on lights at night if they have to get out of bed. They may also wait, sitting at the edge of the bed, before getting up in the dark to allow themselves to awaken more fully and for their eyes to adjust to the darkened room. They should be advised to use lights that come on automatically and to have emergency lighting inside and outside the house in case of a power failure. Patients may need to learn how to plan to move around places with busy visual environments such as shopping malls and grocery stores. For some patients, moving in busy environments may require the use of some type of assistive device, such as a shopping cart or a cane, but for many patients with BVL, no assistive devices are needed after the patient becomes comfortable walking in that environment.



EVIDENCE THAT EXERCISE FACILITATES RECOVERY

Support for the use of exercises to improve physical function in patients with bilateral vestibulopathy

was based originally on the result of studies of nonhuman primates (Igarashi et al).¹⁹ Evidence now exists in people with BVH that supports the use of specific exercises (adaptation, substitution, balance and gait) to decrease subjective complaints, improve visual acuity during head movement, and improve postural stability during functional activities.

TREATMENT

The treatment approach for patients with complete loss of vestibular function involves the combined use of gaze stabilization exercises and exercises that foster the substitution of visual and somatosensory information to improve postural stability and the development of compensatory strategies that can be used in situations where balance is stressed maximally. This approach is also used in the treatment of patients with unilateral vestibular hypofunction.

Future Treatment

Various devices and technologies, such as auditory cues, tactile cues applied to the torso, and stimulation of the tongue have been employed in an attempt to replace the lost vestibular function. Although these devices have shown potential benefits, the devices are solely focused on improving postural control, not on improving gaze stability during head movements. With the success of cochlear implants, several labs have been working on developing a vestibular prosthesis. The basic design is that sensors will detect and measure the directions of rotation and then electrically stimulate the appropriate ampullary nerves. The majority of the work to date has been performed in bilaterally vestibular-deficient chinchillas and monkeys. Della Santina and colleagues have demonstrated partial restoration of the angular VOR to horizontal, as well as RALP (right-anterior, left-posterior) and LARP (left-anterior, right-posterior) rotations with unilateral implantation of the vestibular prosthesis in chinchillas treated with gentamicin.²⁰ In addition to the angular VOR improvements, they noted improved postural stability in the animals as well. Similar improvements in angular VOR responses were observed in rhesus monkeys with bilateral vestibular deficits after unilateral implantation of the vestibular prosthesis.²¹ Even though the responses were not normal, the responses were strong and had similar dynamics to normal animals, and the implanted animals had not undergone any adaptation treatments with the vestibular prosthesis. These studies bode well for future

FUTURE TREATMENT AIMS TO REPLACE LOST VESTIBULAR FUNCTION USING DEVICES FOCUSED ON IMPROVING POSTURAL CONTROL.

implantation of a vestibular prosthesis in humans with bilateral vestibular hypofunction, which may, in combination with rehabilitation, lead to improved function and decreased disability.

In 2021, a study conducted by Margaret Chow and colleagues at Johns Hopkins Hospital was published, focusing on the impact of unilateral vestibular implantation in eight patients with bilateral vestibular hypofunction. These cases, varying from 2 to 23 years, were attributed to either ototoxicity (7 patients) or idiopathic/unknown causes (1 patient). Observations made six months and one-year post-implantation indicated general improvements in posture, gait, and quality of life. However, implantation resulted in reduced hearing in implanted ear apart from one patient.²³

SUMMARY

Studies have shown that the worse disability is in part dependent on the number of comorbidities the patient has as well as greater initial intensity of disequilibrium, lower balance confidence, and poor fall risk scores. Patients with bilateral vestibular problems can be expected to return to many activities but will continue to have problems in certain areas. They will be able to ambulate without the use of a cane or walker, at least when they are in well-lighted environments.²² Patients should be able to return to work, but it may be necessary to find a different occupation. Patients often report difficulty driving, particularly in the rain or snow or at night or on high-speed roads, but actual driving habits and accidents do not differ from subjects with normal vestibular function.^{23,24} Treatment approaches include increasing the function of the remaining vestibular system, inducing the substitution of alternative mechanisms to maintain gaze stability and postural stability during head movements, and modifications of the home and working environment for safety.

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