Bilateral vestibulopathy treatment: update and future directions

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Bilateral vestibulopathy or BVH is a disorder of both labyrinths and/or vestibular nerves which have various etiologies. BVH is most often a chronic condition in which patients can suffer from blurred vision (oscillopsia), impaired spatial orientation and postural instability. Moreover, there is no continuing distressing vertigo, spontaneous nystagmus, or postural falls, which are typical signs of a vestibular tone imbalance caused by acute unilateral lesions. Those symptoms lead to an important decrease in physical activity, social functioning and vitality that dramatically impact the patients' quality of life. The treatment options for various forms of BVH could be one of the following four lines of treatment: (a) Preventive treatment through prevention of ototoxicity, (b) therapeutic treatment through medical treatment of the causative underlying disease, (c) rehabilitative treatment through the vestibular rehabilitation therapy, (d) future directions through sensory substitution devices (balance prostheses technology). The prognosis of BVH is poor and more than 80% of the patients do not improve. The aim of this study was to discuss the update and the future directions in the treatment of the bilateral vestibular hypofunction (BVH).

Keywords:
bilateral vestibulopathy, dizziness, oscillopsia, sensory substitution devices

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Background

Bilateral vestibulopathy or bilateral vestibular hypofunction (BVH) is a disorder of both labyrinths and/or vestibular nerves which has various etiologies. It was first described by Dandy (1941) in patients who had undergone bilateral vestibular neurectomy. Zingler and colleagues reported that BVH accounts for ~1–2% of all individuals undergoing electronystagmography studies.

Generally patients with BVH are first referred not only for assessment of dizziness and disequilibrium, but also for examination of ocular motor disorders, ataxia, or hearing loss, conditions in which BVH is often not suspected before investigation [1]. A chronic bilateral loss of vestibular function is surprisingly well tolerated. This is due to the fact that spatial orientation, posture, and eye movements are mediated by redundant multisensory processes, which can in part compensate for each other's deficiencies. Moreover, there is no continuing distressing vertigo, spontaneous nystagmus, or postural falls, which are typical signs of a vestibular tone imbalance caused by acute unilateral lesions [2].

The key symptoms are oscillopsia during locomotion or head movements and unsteadiness particularly in the dark. The diagnosis is made with the simple bedside test for defective vestibulo-ocular reflex during rapid, passive head turns (head thrust test) [3]. It is confirmed by the absence of nystagmic reaction to both caloric and rotatory chair testing. The most frequent etiologies include ototoxicity, autoimmune disorders, meningitis, neuropathies, sequential vestibular neuritis, cerebellar degeneration, and tumors; idiopathic BVF is found in more than 20% of the patients [4].

Subjective symptoms in the acute stage tend to improve with time by processes of somatosensory and visual substitution of vestibular function. Vestibular rehabilitation is supportive of this improvement, patient response to physical therapy with gait and balance training is quite positive. The spontaneous recovery of patients with BVH is relatively rare and incomplete. A permanent loss of vestibular function is the most frequent form; however, patients with BVH remain largely asymptomatic until confronted with high-frequency motion conditions or situations where proprioceptors or vision cannot replace the deficient vestibular system [5].

Management

The treatment options for various forms of BVH could be one of the following four lines of treatment:

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(a) Preventive treatment through prevention of ototoxicity,
(b) Therapeutic treatment through medical treatment of the causative underlying disease,
(c) Rehabilitative treatment through the vestibular rehabilitation therapy (VRT) [6],
(d) Future directions through sensory substitution devices (balance prostheses technology) [7].

Preventive treatment

Furman and Cass (2003) reported that prevention is the best management for ototoxicity. Aminoglycoside therapy should be used only if strictly indicated and then only in a once-daily dose. Plasma levels should also be monitored. Patients with renal insufficiency, advanced age, prior use of ototoxic drugs, high serum levels of ototoxic drugs, pre-existing sensorineural hearing loss, and a medical course greater than 14 days are at particular risk. Some patients may have a genetic predisposition to aminoglycoside ototoxicity but there is currently no clinically available test for this condition [6]. Ototoxic antibiotics should not be combined with other ototoxic substances, such as loop diuretics, as this can greatly increase inner-ear damage. Careful monitoring of the hearing and vestibular function is necessary during treatment. However, the physician must remain vigilant, as the ototoxic effects of gentamycin have a delayed onset, often appearing only after days or weeks [8].

Hain and colleagues suggested that if the damage has already been done, then the focus of preventive treatment is upon avoidance of vestibular suppressants and ototoxins (Table 1). Vestibular suppressants typically will cause temporary worsening of dizziness or hearing symptoms while ototoxins will cause permanent or temporary worsening. These medications need not be avoided completely, but reasonable judgment should be exercised. Medications that cause only temporary unsteadiness (such as meclizine), may still be useful in some situations. Medications that are ototoxic (such as gentamycin), may still be useful in cases of BVH when there is no reasonable alternative or when the damage done is already so extensive that there is nothing more to lose. Certain bacteria, such as *methicillin-resistant Staphylococcus aureus* are so difficult to treat that ototoxic medications may be required to save a person's life.

Therapeutic treatment

Treatment of the causative underlying disease is important in BVH. Medical treatment and recovery are possible by treating immune-mediated inner ear disease, which are diagnosed too infrequently [10]. Although controlled prospective studies are lacking, immune treatment is theoretically expedient, only if there are clinical signs of a systemic autoimmune disease or if antibodies against inner ear structures are detected [3].

Moscicki (1994) suggested that the treatment should be initiated with corticosteroids as early as possible, for example, prednisone in doses of 60 mg daily for adults and 1 mg/kg body weight daily for children for a period of at least 30 days. Improvement may not be symmetrical, usually the most recently affected ear responds best. If the response is inadequate or relapses occur, addition of cyclophosphamide or azathioprine or methotrexate may be considered as well as plasmapheresis or high-dose intravenous g-globulins. In Cogan's syndrome, higher doses of steroids (1 g intravenously for 5 days, then tapered for several weeks) are recommended [6].

Rehabilitative treatment

The use of VRT to treat patients with BVH has been around for many years. It is a specific form of physical therapy designed to promote mechanisms of central adaptation and compensation for a wide variety of balance disorders. The goals of VRT are to improve gaze and postural stability, minimize falls, and decrease patient's sense of disequilibrium and off-balance [11].

Significant improvements are reported in dizziness and balance after vestibular rehabilitation. The neurophysiologic basis of these changes is believed to be due to adaptation, habituation, and sensory substitution or reweighting of the central nervous system [12]. The vestibular system, and in particular

Table 1 Vestibular suppressants and ototoxins [9]

Vestibular suppressants	Ototoxins
Antihistamines such as meclizine (Antivert), dramamine and phenergan	Cis-Platinum (a chemotherapy drug) and other platinum based drugs.
Antidepressants such as amitriptyline, especially tricyclic type antidepressants	Gentamycin and other 'mycin' antibiotics, including large doses of erythromycin (although this is actually in a different group than gentamycin)
Aspirin or NSAIDs (drugs like ibuprofen and naproxen) in large doses	Furosemide (Lasix) and ethacrynic acid (Edecrin) loop diuretics
Diazepam (Valium), alprazolam (Xanax), lorazepam (Ativan), clonazepam (Klonopin) and related drugs in the benzodiazepine family	Quinine and related drugs (they usually have a 'quin' in their name)
Verapamil and other calcium channel blockers	

the vestibulo-ocular reflex (VOR), is highly adaptable or plastic; its adaptive capability is important during development and maturation, and in response to disease and injury. The potential for adaptation can be used in patients with vestibular hypofunction as a mechanism to induce recovery. The signal for inducting adaptation is the movement of an image across the retina while you are trying to keep the image in focus. This creates an error signal (i.e. retinal slip) which the brain attempts to minimize by increasing the gain of the VOR and function improves [7].

In habituation exercises, the provoking position or stimuli is repeated at regular intervals until the person no longer responds to the stimuli. The earliest vestibular rehabilitation exercises (Cawthorne-Cooksey exercises) were believed to be a form of habituation [13]. Another mechanism of recovery is the sensory reweighting or substitution of alternative strategies for the lost function. Sensory reweighting is the brain's ability to change the relative reliance on a specific sensory modality for orientation depending on environment, task, or pathology [7].

Telian and Shepard (1996) suggested that the starting point in developing an exercise program is the thorough evaluation of the patient. Evaluation must take into consideration not only vestibular system functions, but also visual, somatosensory, motor, and biomechanical factors. A customized VRT program adapted to suit the specific needs of the individual became widely accepted in the 1990s and has become the primary modality of treatment for a large number of patients with dizziness. Treatment can be delivered in the form of supervised outpatient exercises or home exercise program. The average duration of a VRT program ranges from 4 to 10 weeks. One important consideration in designing a treatment program for patients with BVH is whether there is any remaining vestibular function, as the program would involve exercises to enhance remaining vestibular function, as well as substitution strategies to replace the lost vestibular function to improve gaze and postural stability [7].

Traditional vestibular rehabilitation therapy

Exercises to promote alternative strategies for gaze stability

Strategies to improve gaze stability include: (a) potentiation of the cervical-ocular reflex, where receptors in the muscles and joint facets in the upper cervical spine region project to the contralateral vestibular nucleus, producing a compensatory slow phase eye movement. This compensatory eye movement is opposite to the direction of head movement during low-frequency, brief head movements and complements the VOR [14]; (b) modification of saccades either by decreasing the amplitude of saccades or the use of corrective saccades during combined eye and head movement to maintain gaze stability on a visual target; (c) increased smooth pursuit eye movement; (d) central preprogramming of eye movement, this is an effective compensatory eye movement strategy to maintain gaze stability if the task is predictable. Central programming of eye movements is based on the prediction or anticipation of intended motor behavior [7]. All the above substitution strategies are, however, only effective for low frequency head movement (<1 Hz); (e) restriction of head movements to fixate gaze.

Gans (1996) and Herdman and Clendaniel (2007) suggested the following exercises to improve gaze stability:

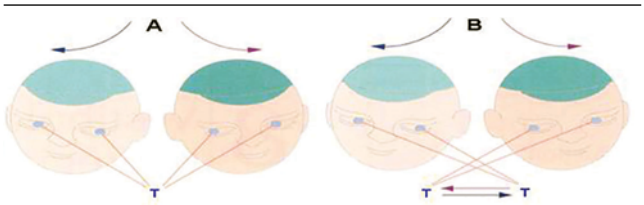
(a) VORX1 and VORX2 viewing exercises to improve the remaining vestibular function and central preprogramming of eye movements;
(b) The exercise involving active eye–head movements between two targets helps to facilitate the use of saccadic or smooth pursuit strategies and central preprogramming;
(c) Imaginary target exercise helps to facilitate the central preprogramming of eye movements (Fig. 1).

Exercises to improve static and dynamic postural stability

Clendaniel (2002) suggested that the initial postural stability exercises for patients with BVF are simple standing balance exercises, with the goal of developing independent stance. As the patient is able to stand without assistance, the exercise is made more challenging by progressively decreasing the base of support, until balance cannot be maintained. The static postural stability exercises can be performed initially with the arms outstretched, then at the side, and finally folded across the chest. In these exercises both visual and somatosensory cues are available to the patient.

Herdman and colleagues (2001) reported that individuals with BVH tend to initially rely solely on visual cues but gradually utilize somatosensory cues

Figure 1



VOR stimulation exercises, VORX1 (a) and VORX2 (b), (T = target) (quoted from Clendaniel and colleagues).

for postural stability. If the examination of the patient reveals that they are not using somatosensory cues for postural stability, then the standing balance exercises can be performed with the eyes closed. Eye closure may need to be intermittent at first but is progressed until the patient can perform the exercise for 30 s. In addition, active weight shifting with eyes closed increases somatosensory awareness and awareness of the supporting surface. The postural stability exercises can be performed with eyes open while standing on a compliant surface, such as a foam pad, to perturb the somatosensory inputs. Altering visual and somatosensory input to optimally challenge the individual should only be performed with the necessary safety measures to prevent falls.

Patients with bilateral vestibular failure (BVF) also tend to walk with a wide base of support. Having these patients walk with a decreased step width, ultimately ambulating heel to toe and changes in direction can challenge their dynamic postural stability. An obstacle course or functional tasks that require positional changes, movement, and changing visual or somatosensory cues are also beneficial by forcing the patient to adapt to changing environmental conditions and to shift between the available sensory cues [15].

Clendaniel (2002) suggested that patients with BVH should be instructed in behavioral and environmental modifications to optimize function and reduce the risk of falling. Environmental modifications may include adequate lighting (including emergency lighting in case of a power failure), removal of thick carpeting and area rugs, nonskid floors, pathways clear of obstructions, stable tables and chairs, reorganization of cabinets and shelves to minimize bending and reaching, and stairs with railings. Moreover, it includes using cane or walker in complex environment to avoid falls.

Behavioral modifications may include learning to plan movements, avoiding hurried movements, sitting to work, visual fixation on distant objects for stability, light touch on objects for balance, restricting rapid head movements and walking in crowds. At its extreme, some patients may modify their behavior to avoid situations where visual or somatosensory cues are diminished such as going out at night [15].

Instrumental vestibular rehabilitation therapy (biofeedback technology)

Maintaining balance is a complex task accomplished by the brain through the fusion and interpretation of sensory information. When sensory information from vestibular, somatosensory, and visual systems are not accurate and/or adequate, balance will be compromised [16]. Although, in many cases, the loss of peripheral sensory information is not curable or reversible, the brain can compensate for the loss of sensory information by relying more on the other sensory channels [17].

Examples of instrumental rehabilitation techniques for vestibular dysfunction include use of biofeedback (BF) technology and virtual reality technology. The purpose of BF systems is to provide additional sensory information about body equilibrium to the brain [18]. Virtual reality is a method of treating people with inappropriate visual dependence.

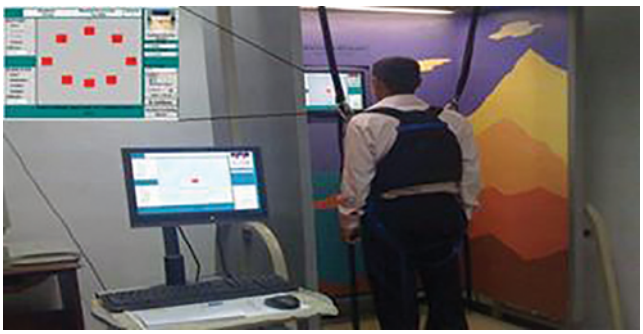
In the last few years, different sensors, encoding algorithms, and information restitution devices have been combined to develop promising BF systems for postural control. The major design goals were focused on portability, usability, economy, and effectiveness in balance improvements [19]. The development of these BF systems has been facilitated by the availability of lightweight, miniaturized, and economical sensors such as accelerometers, inclinometers, and gyroscopes [5].

Currently research is being conducted to determine whether individuals with BVH may benefit from some form of sensory stimulation to replace the absent vestibular signals. Although this work is in its infancy, the preliminary results are promising [7]. Danilov and colleagues (2006); Danilov and colleagues (2007); Vuillerme and Cuisinier (2008) studies have demonstrated the efficacy of the BF technology to replace the absent vestibular signals. The purpose of BF devices and training programs for postural instability is to substitute missing sensory information for balance and provide additional sensory information about body equilibrium to the brain. The idea of BF systems is based on encoding of head and/or body position and movement into visual (visual BF), auditory (auditory BF) cues delivered to the patient to augment postural stability [7].

Visual biofeedback forceplate training (posturography training)

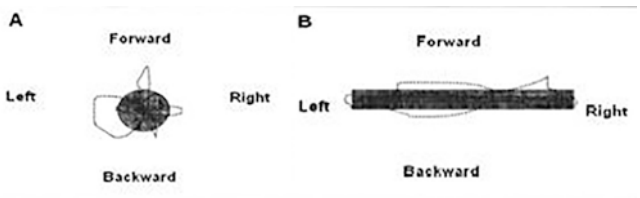
Recent advances in technology have resulted in the commercial availability of numerous force platform systems for the retraining of balance function in patients with postural instability, including patients with BVH. These systems are designed to provide visual or auditory BF to patients regarding the locus of their center of gravity (COG), as well as training protocols to enhance stance symmetry, steadiness, and dynamic stability [20]. Typical force platform BF systems consist of at least two force plates to allow the weight on each foot to be determined, a computer and monitor to allow visualization of the COG, and software that provides training protocols and data

Figure 2



Visual biofeedback training screen.

Figure 3



Steadiness training with central target: (a) in this task, the subject is asked to maintain the computer cursor inside the shaded circle. (b) in this task, the subject is asked to shift weight repetitively from left to right while maintaining the cursor within the shaded rectangle (quoted from Nichols).

analysis capabilities (Fig. 2). Some units allow auditory feedback in addition to the visual feedback in response to errors in performance. This procedure is a relatively new rehabilitative tool and little outcome information is available about its effectiveness [20].

Nichols (1997) reported that three types of balance measures are most commonly used by force platform systems to evaluate balance function and patient progress related to balance ability: steadiness, symmetry, and dynamic stability. Steadiness refers to the ability to maintain a given posture with minimal extraneous movement (sway). The term symmetry is used to describe equal weight distribution between the weight-bearing components (e.g. the feet in a standing position, the buttocks in a sitting position), and dynamic stability is the ability to move within a given posture without loss of balance [21]. All of these components of balance (steadiness, symmetry, and dynamic stability) have been found to be disturbed in the vestibulopathic patients. Balance testing of patients with BVH has revealed a greater amount of postural sway during static stance, asymmetry, and a decreased ability to move within a weight-bearing posture without loss of balance [21].

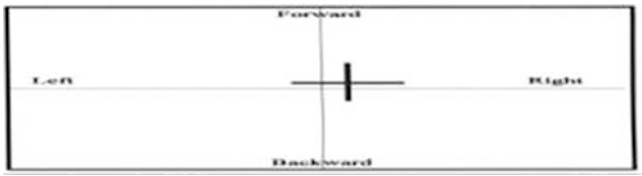
Balance retraining with visual BF can address each of the components of function described (steadiness, symmetry, and dynamic stability). Postural steadiness can be addressed through activities that require maintenance of the COG, usually depicted by the cursor on a computer screen, within a narrow target or within a narrow range, designated by a shaded area on the screen, as weight is transferred from one target to the next (Fig. 3) [21].

Most studies that have evaluated the use of postural BF have emphasized stance symmetry in their training protocols. Postural symmetry can be addressed by maintaining the COG in midline by having subjects maintain a cursor in the center of a target on the computer screen (Fig. 4), or by providing visual information regarding the percentage of weight on each foot. The patient can be asked to perform various activities while maintaining equal weight distribution, such as coming to a standing position with equal weight distribution, reaching to the side and returning to a symmetrical stance, and stride standing and stepping [22]. Furthermore, dynamic stability training, involving weight shifting to successive targets, has also been found to increase stance symmetry. Studies of Dettmann and colleagues (1987); Nichols and colleagues (1995); Liston and Brouwer (1996) showed increased stance symmetry following training, and in those studies that had a control group, the increase in symmetry was greater in the subjects who received the BF training than in the control subjects who received traditional physical therapy.

The training of dynamic stability, referring to movement within the limits of stability, is most commonly done by having subjects shift weight along the anteroposterior or mediolateral plane or to selected targets displayed on a computer screen. Two protocols have been described most consistently in the literature [23]. One protocol involves a central target encircled by a series of targets at 45° angles (Fig. 5a). The subject's task is to shift his or her weight forward to a lit target and back to the central target within a specified period of time, typically 7–10 s, before the next target is illuminated. The transition time (time to move the COG from the starting position to the target), the sway path (cumulative distance covered), the sway error (accuracy of the weight shift from the central target to the peripheral target, and peripheral sway area (sway magnitude once the target is reached) are units used to evaluate patient performance [21].

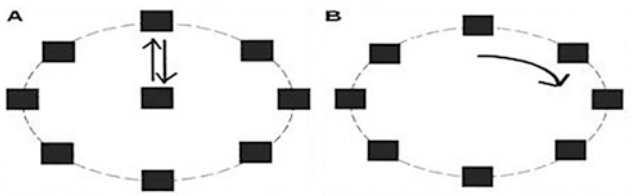
The other protocol involves shifting weight around a series of successive targets oriented in a circle at 50–75% of the individual's limits of stability (Fig. 5b); again, transition time, sway path, sway error, and peripheral sway area are the units used to evaluate subject

Figure 4



Symmetry training with central target: in this task, the subject is asked to maintain the computer cursor (+) in the center of the computer screen as marked by the cross here. The sway path may be depicted on the screen (not illustrated) (quoted from Nichols and colleagues).

Figure 5



(a) Dynamic stability from center to target; (b) dynamic stability to successive targets. This can be done in either a clockwise or counterclockwise manner. Transition time, path sway, and distance error can be calculated (quoted from Nichols).

performance. This type of training has been found to decrease the magnitude of each of these variables, which indicates an increased accuracy of the weight shift. In addition, subjects with postural instability have been able to extend their limits of stability with dynamic stability training. Expanding these limits should decrease the likelihood of falling [21]. Furthermore, this type of training often addresses more than one balance component; activities that encourage stance symmetry also require minimal postural sway for the patient to be successful, and activities that involve weight shifting for dynamic stability also often address postural sway and symmetry in order for the target to be reached quickly and accurately [24].

Treatment strategies involving visual feedback exercises require training over repeated sessions, typically two sessions are given per week over several weeks; therefore, a practice effect is introduced which must be taken into account when evaluating any rehabilitative outcome of therapy [25]. So, data is needed to assess if body sway is reduced after a series of practice sessions using visual feedback for people without any balance disorders. The normative training effect must be separated from any rehabilitative effect. This information must be considered when observing the performance of patients, and before assessment of their progress is made [25].

This procedure seems unlikely to promote neuroplasticity or adaptation because it is too short, but it might assist individuals in forming internal models of their body and the outside world. Forming and recalibrating internal models is certainly a worthwhile endeavor, critical to recovery from lesions. It seems likely to occur in time whether or not a device like this is available, but the progress of revising an internal model might be accelerated through guided practice [20]. Moore and Woollacott (1993) reported that the visual feedback is supposed to be used to match and recalibrate proprioceptive sensory information or input that may be impaired.

NeuroCom International Inc. (2002) suggested that to maximize the benefit of a rehabilitation program, the prescribed exercises must be customized to target the specific impairments of the individual patient to resolve the underlying impairments and functional limitations. A generic approach to balance treatment will not be as effective as an approach that singles out the underlying impairments, and may not improve function at all. Rehabilitation outcomes are enhanced by motivating patients to exercise as intensively as possible. To keep the patient challenged and maximize learning, the difficulty level of the focused exercises can be progressed by increasing the limits of stability and the pace (time allowed per weight shift) to challenge the subjects' weight-shifting abilities as their balance improved over the course of rehabilitation program [26].

Immediate feedback information can help clinicians adjust the exercise task difficulty to match the maximum capabilities of the patient. An overly difficult task results in a very low success rate, while a very high success rate indicates that the task is insufficiently challenging. Furthermore, structured exercises and objective feedback provide the additional benefit of maintaining the patient's motivation and performance accuracy while reducing the level of clinician supervision [25].

Results of a recent study by Corna and colleagues. (2003) comparing posturography training to Cawthorne-Cooksey exercises indicate that after only 1 week of twice-daily supervised therapy, the group with posturography training showed significant improvements in objective and subjective measures that were maintained for 1 month post-treatment. However, self-perceived handicap, clinical balance scores, and postural sway improved significantly more for patients undergoing rehabilitation with the support surface translations. The investigators conclude that although both treatments are beneficial, training on a moving platform is more effective particularly for improving postural control. Further research is needed to determine the optimal support surface stimulus needed to improve postural control [27].

Sensory substitution devices

Nonimplanted vestibular prostheses (vibro-tactile stimulation)

Recent studies of Danilov and colleagues (2006); Danilov and colleagues (2007); Vuillerme and colleagues (2008) have demonstrated the efficacy of balance prostheses which encode head and/or body position and movement into tactile cues delivered to the trunk, tongue and the cranium to augment balance in patients with BVH. These devices translate information normally sensed by the vestibular system into a proprioceptive cue that is then integrated with the remaining visual and somatosensory inputs to augment postural stability. The vestibular replacement prostheses are available in the form of: body fixed vibrotactile stimulation, head fixed vibrotactile stimulation (tongue, cranium) and implanted vestibular evoked stimulation. Head mounted vibrotactile stimulation leads to improved truncal stability and improves subjective sense of verticality.

Studies of Vuillerme and Cuisinier (2008) preferred the tongue as a substrate for the electrotactile stimulation site because of its neurophysiologic characteristics, it has dense mechanoreceptive innervations and large somatosensory cortical representation, and it can convey higher-resolution information than the skin can. In addition, due to the excellent conductivity offered by the saliva, electrotactile stimulation of the tongue can be applied with much lower voltage and current than is required for the skin.

Wicab Inc. has developed the BrainPort balance device which transmits information about the patient's head position through electrotactile stimulation of the tongue. Head position data (artificially sensed by a micro-electro mechanical system accelerometer) serve as the input signals for the BrainPort balance device. Using this data, the device generates a small pattern of stimulation on the tongue that relates to head position in real time (Fig. 6). This pattern of stimulation moves forward, backward, and laterally on the tongue in direct response to head movements. Users of the device were trained to use this stimulation to adjust their position to maintain their balance. A specialized set of training procedures were developed to serve as the course of therapy with the BrainPort balance device. We hypothesized that with training, the information presented on the tongue from the BrainPort balance device could be acquired, retained, and transferred by the user to improve both static and dynamic balance [28].

Barron Associates Inc. has developed a prototype head-mounted vestibular improvement prosthesis, which provides information about head motion to vestibulopathic patients as a balance aid. The prototype vestibular improvement prosthesis included a Newton's Apple unit, which provided the sensing of head motion/orientation, and four tactors capable of exciting the scalp (through a low-amplitude vibration stimulus) in such a manner as to provide the wearer with a sense of head tilt independent of their perceptions. Newton's Apple and tactors were mounted on the head along the same transverse plane using a custom elastic headband (Fig. 7) [29].

Vestibular implant

Vestibular implants are a promising tool to treat bilateral vestibular loss since neither medication nor rehabilitation is sufficient. Electrodes are implanted in the three semicircular canals, and it has been shown that electrical stimulation of those canals can partially restore vestibular functionality [30,31]. The architecture of the vestibular implant is very reminiscent of that of cochlear implants (Fig. 8); it has an external part that registers head movements and which can be secured to the head, and an internal part that connects to the nerve ends. The external component contains gyroscopes and accelerometers, and a microprocessor that digests the measurements and transmits results to the internal part which has electrodes that are connected to the vestibular nerve; briefly, gyroscopes measure movement, and they give the signal to a processor, which converts it into the desired digital signal. The desired vestibular branch is then stimulated [31].

Figure 6

Sensory coding schemes for the tongue display unit (TDU) (b) as a function of the head orientation with respect to gravitational vertical (a). (1) Neutral, (2) right-side-tilted, (3) extended, (4) left-side-tilted and (5) flexed head postures (quoted from Vuillerme and Cuisinier).

Figure 7

Close-up view of headband housing tactors (small arrow) and Newton's Apple (large arrow) (quoted from Goebel and colleagues).

Figure 8

Design of the vestibular implant (receiver/stimulator and electrode); the implant consists of three channels with three electrodes each (nine electrodes total) (quoted from Rubinstein and colleagues).

A vestibular implant bypasses the vestibular end organs to directly stimulate the vestibular nerve much as a cochlear implant bypasses the cochlea to stimulate the cochlear nerve directly. Before development of implant prototypes, it was known that electrical stimulation of the vestibular nerve could induce the vestibulo-ocular reflex [32] and modulate balance [31]. Human studies have even shown that balance abilities can be enhanced during platform perturbations using transcutaneous stimulation of vestibular afferents (galvanic vestibular stimulation) [33]. The goal of vestibular implantation is to provide vestibular functionality and/or reduce symptomatology by programmed or sensory-mediated stimulation of the vestibular nerve.

The idea of the vestibular Implant is currently investigated by research groups in Europe and the USA (the Geneva-Maastricht groups). Research both from animal and human studies have demonstrated that electrical stimulation is an effective means to activate the vestibular system [34]. The two groups, who are working together, used a slightly different approach: in Maastricht the electrodes were attached within the semicircular channels of the vestibular system, while in Geneva the electrodes were attached outside the vestibular system. All used implants that are prototypes developed by Medel, a well-known manufacturer of cochlear implants [32].

The first vestibular implants, developed to replace the function of the vestibular system, were recently installed in three patients at two facilities in Europe. The current implants only restore the function of the semicircular canals, which are thought to be the most important and are technically easier to restore by putting electrodes into the ampulla of the canals, close to the branches of the vestibular nerve [32].

Guyot and colleagues (2011) reported that the vestibular implant is being developed primarily for patients with BVH. It is still too early to tell whether the implants work as conceived, something that will only become clear in the coming period after the implant has been further fine-tuned. Previous animal studies, however, were successful in restoring balance with similar implants, so results of these first implantations in humans are eagerly anticipated.

BVH, for example, shows that as a result of the disease, an individual's physical activity, social life, vitality, and quality of life decreases. This is the result of the fact that gaze stabilization, balance, and spatial orientation are not automatically processed anymore. The next steps depend on the results from the tests. For instance, determining the right stimulation paradigm, measuring adaptation of the brain to the vestibular implant, determining the effect of the implant on balance tests, and the effect on the quality of life [32].

Prognosis

BVF is a condition that realistically often causes some permanent disability as the recovery from this disorder is seldom to a normal status. A deficit in balance control will remain even though the somatosensory and the visual systems can provide some compensation for the vestibular loss. However, each of these systems has its own velocity/acceleration and frequency limitations and, therefore, incompletely substitutes for BVF in certain environments. Recovery of vestibular deficit and hearing loss is possible in postmeningitis cases due to a serious nonsuppurative labyrinthitis [1].

Vibert and colleagues (1995) reported that partial recovery has been described in more than 50% of patients with simultaneous or sequential idiopathic bilateral vestibulopathy. In patients with gentamycin induced ototoxicity, the symptoms generally peak at 3 months from the last dose of gentamycin. In the long run (5 years), however, most patients are substantially better. There are multiple reasons why people get better. First of all, there is evidence that the damaged vestibular hair cells in the inner ear can regenerate, although the extent to which this occurs and the degree to which they are functional is not presently clear. Second of all, the brain rewires itself to adapt to the new situation (plasticity and substitution), also people adjust their way in doing things to their situation.

Finally, recent experimental work suggests that high frequency vestibulo-ocular response to rotation recover through a pathway that does not require vestibular input [7]. Thus in time, oscillopsia should improve no

matter how severe the vestibular deficit is [7]. Hain and colleagues (2013) reported that recovery is related to various factors, including severity of lesion and prognosis based on the amount of damage done initially, modified by other factors such as age, and other medical problems.

Hain and colleagues (2013) noticed that rotatory chair testing done at 6 months following onset (or later) helps to establish prognosis by dividing individuals into three categories. Individuals with mild abnormalities on rotatory testing are nearly always subjectively normal at 1 year. Individuals with moderate vestibular loss are usually able to continue to work productively, with some modifications in their behavior. For example, 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.

In situations where there is complete or near-complete loss of vestibular function, vision and balance usually remain impaired permanently; however, most individuals do return to work, especially if their job does not require good head/eye coordination or balance. Frequently, job modification or accommodation occurs. For example, it would not be safe to continue as a truck driver, construction worker, or a roofer. A job where you work at a desk is usually a good choice [9].

Reading is generally more difficult than for persons with normal vestibular systems, but quite feasible, as the head can be steadied during reading. Many people with bilateral vestibulopathy complain of a mild confusion or 'brain fog,' which is attributed to the increased attention needed to maintain balance and vision. This reduces the amount of attention that is available for other thinking tasks [8]. While crutches, canes, walkers and wheelchairs may be necessary in the first 3 months, these appliances are rarely needed by 1 year. After 20 years, most patients have returned to near-normal for their age. To some extent this return to 'normal' is related to the aging of the patient's peers, since vestibular function normally declines with age. The long-term prognosis has not been sufficiently studied [9].

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Conflicts of interest
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