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Bilateral Idiopathic Loss of Peripheral Vestibular Function with Normal Hearing*

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From 1982 to 1993, 52 electronystagmograms (ENG) revealed an absent nystagmic reaction on both caloric (44–30 and 10°C water irrigation) and rotatory pendular testing (0.05 Hz/peak velocity 60°/s), which represents 0.6% of all ENG performed during this period. Among these examinations, 14 patients (27%) presented a bilateral idiopathic loss of vestibular function (BILVF) with normal hearing and without associated neurological symptoms. Two different groups emerged: one group with simultaneous onset of BILVF (10 patients), with sudden imbalance and disequilibrium, worse in darkness, with an absence of bilateral caloric and pendular response. The other group (4 patients) was characterized by sequential onset of BILVF. These patients experienced several episodes of acute vertigo with persistent imbalance; caloric and pendular responses showed unilateral, then contralateral loss with or without recovery of function. Eleven were controlled with a follow-up from 1 to 7 years. Patients of both groups remained permanently or episodically symptomatic, but only 4 patients complained of persistent oscillopsia. Viral infections, systemic diseases (hypertension, hypothyroidism, asthma), immune reactions (vaccination) and toxic factors (herbicide exposure) may play a role in the etiology of this rare bilateral peripheral vestibulopathy. *Key words*: *bilateral vestibular areflexia, oscillopsia, disequilibrium, bilateral vestibular neuritis.*

INTRODUCTION

Bilateral loss of peripheral vestibular function is a rare and often unexpected finding in patients complaining of dizziness or disequilibrium. In 1941, Dandy described two specific secondary symptoms following bilateral vestibular neurectomies performed on patients with bilateral Meniere's disease (1). The first symptom was oscillopsia, a visual distortion like a jumbling image, occurring either during motion or with head movements. A severe imbalance was observed during darkness as the second symptom. Bilateral loss of vestibular function is for this reason also called "Dandy Syndrome". Few studies in the literature have been reported regarding this particular otoneurologic condition (2–6).

Among all the electronystagmograms (ENG) performed between 1982 and 1993 in the neuro-otological laboratories of Berne and Geneva, 52 (0.6%) revealed an absence of bilateral nystagmic reaction with both caloric (44–30 and 10°C water irrigation) and rotatory pendular testing (0.05 Hz/peak velocity 60°/s). The purpose of the present paper was to describe in particular the 14 patients (27%) which presented a bilateral *idiopathic* loss of vestibular function (BILVF) with normal hearing and without associated neurological symptoms.

MATERIAL AND METHODS

From 1982 to 1993, 8,396 ENG were performed.† Review of the records revealed 52 ENG (0.6%) with a bilateral loss of vestibular function. This condition was defined in our series as a complete loss of response to both caloric (44–30–10°C water irrigation) and rotatory pendular testing (0.05 Hz/peak velocity 60°/s). The study group included 39 males and 13 females (aged from 3 to 82 years). The etiologies of the 52 cases of bilateral loss of vestibular function are listed in Table I.

Among these patients, 14 (27%) presented a BILVF with normal hearing and without associated neurological symptoms. They underwent a neuro-otological examination including a clinical vestibular examination, a classic pure tone audiogram, stapedial reflex measurements, brainstem auditory evoked potentials, and electronystagmography.

Electronystagmography consisted of recording the spontaneous nystagmus with (light) and without (darkness) visual fixation; the positional nystagmus with the head in hyperextension, then turned to the right and to the left (positions of Rose); the optokinetic nystagmus at speeds of 25, 50 and 75°/s (rotation

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† 8008 ENG from the Neuro-otology Unit of the Department of Otolaryngology, Head & Neck Surgery, Cantonal University Hospital of Geneva were performed from 1982 to 1992 and 388 ENG were performed in 1993, in the University clinic of ENT, Head & Neck Surgery, Inselspital, Berne.

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Table I. *Etiology of all patients showing a bilateral vestibular loss at the neuro-otological examination (ENG) (n = 52)*

Idiopathic	25	(48%)
Ototoxicity (Garamycine)	11	
Brainstem tumors (bilateral acoustic neurinoma, lymphoma metastasis)	2	
Heredo-degenerative diseases (Usher, Friedreich, Charcot-Marie)	5	
Meningitis	2	
Bilateral labyrinthitis	2	
Bilateral temporal bone fracture	2	
Bilateral Meniere disease	1	
AIDS	1	
Bilateral inner ear fistula (Cholesteatoma)	1	

to left and right) with whole retinal field stimulation. This was followed by an examination of smooth pursuit, a rotatory pendular test (undamped rotation of 360° in 20 s; sinusoïdal frequency of 0.05 Hz with a peak velocity of 60°/s) with (light) and without (darkness) visual fixation suppression, and a bithermic caloric test with recordings of nystagmus duration after irrigation of each ear for 20 s with 20 cm^3 of water at 44, 30 and 10°C. The corneoretinal potentials were recorded for all examinations simultaneously from both eyes with horizontal and vertical leads. Details of the recording techniques used for ENG were reported previously (7).

The bilateral peripheral vestibular loss was defined as the bilateral absence of nystagmic response to the caloric and rotatory pendular tests. A unilateral absence of nystagmic response to caloric testing as well as an asymmetric nystagmic response (side difference $\geq 40\%$) to the rotatory pendular test were the criteria defining a unilateral peripheral vestibular loss.

RESULTS

Audiological tests were normal in 10 out of the 14 patients presenting a BILVF. Four patients (62 to 82 years old) had a bilateral symmetrical high-frequency sensorineural hearing loss presumably due to aging (presbycusis).

Two different groups with bilateral vestibular loss emerged from the otoneurological history and ENG findings: the loss was simultaneous-bilateral in 10 patients and sequential in 4 patients, i.e. with a unilateral followed by a contralateral loss in a period of months to years. Eleven out of 14 patients (10 males: 1 female; aged from 20 to 82 years (average: 38.2 years), were controlled and serial ENG were performed with a follow-up from 1 to 7 years. Eight patients were in the simultaneous-BILVF group, 3 in the sequential-BILVF group. We describe hereafter 5 representative case reports:

Simultaneous BILVF

Case 1. In July 1985, a 42-year-old man experienced a sudden unsteadiness, worse in darkness, without rotatory vertigo, associated with oscillopsia in motion. There was no history of hearing loss or tinnitus, viral infection or ototoxic drug consumption. Otherwise, the medical history was unremarkable. On examination, caloric and pendular testing showed a bilateral absence of nystagmic response. Examination 7 years later revealed a persistence of imbalance in darkness and inability to ski in fog. The otoneurological examination showed persistent absence of caloric and pendular responses on both sides.

Case 2. In 1990, a 33-year-old man with no previous medical history underwent a sudden acute unsteadiness while walking several minutes after massive respiratory herbicide exposure at his workplace. The imbalance was constant and increased in darkness. There was no hearing loss, tinnitus or oscillopsia, and no rotatory vertigo. Caloric and pendular testing revealed absence of nystagmic response on both sides. An otoneurological check-up 2 years later showed persistent absence of bilateral caloric and pendular responses with normal hearing; the patient complained of a persistent imbalance while walking in darkness without oscillopsia in motion.

Case 3. In February 1990, a 20 year-old man fell with a sudden "feeling of drunkenness" (without alcoholic or other drug consumption) while dancing. Since that time he noted oscillopsia upon motion, difficulties in walking and inability to ride a bicycle in darkness. He had no hearing loss or tinnitus. The otoneurological examination showed normal hearing and absence of bilateral caloric and pendular response. Three years later, the imbalance had improved but persisted in darkness and the patient was continually disturbed by oscillopsia upon motion. The otoneurological control showed persistent absence of caloric response on both sides with a symmetrical nystagmic response to pendular testing.

Sequential BILVF

Case 4. A 62-year-old man treated for a hypothyroidism developed acute vertigo of days' duration. Several years later he complained of a severe imbalance while walking, following an acute episode of rotatory vertigo lasting for several minutes. There was no tinnitus, no ototoxic drug consumption, no oscillopsia. The audiogram revealed a symmetrical high-frequency sensorineural loss. The CT-scan per-

formed was normal. The first examination showed a right loss and a left diminished response to caloric and pendular testing. Five years later, he noted increased imbalance which became incapacitating: the second ENG revealed persistent right areflexia and left hyporeflexia to caloric (10°) testing with normal symmetrical pendular testing. Because of a sudden worsening unsteadiness one year later, a third ENG was performed and showed bilateral absence of nystagmic response to caloric testing. At the last visit, the complaints remained the same; he was unable to walk without a cane. No oscillopsia was seen. The ENG findings showed a left recovery of the caloric response (44°) and a symmetrical nystagmic response on pendular testing.

Case 5. In 1986, a 23-year-old man with no medical history experienced a brief rotatory vertigo episode occurring with head rotations and associated with unsteadiness in darkness. He also noted oscillopsia when walking. There was no hearing loss or tinnitus, and no ototoxic drug exposure. The first ENG showed bilateral absence of caloric and pendular responses. Five years later a new brief rotatory vertigo lasting a few seconds occurred while he was driving his car, followed by a feeling of "drunkenness" for a few hours. The ENG once again showed no vestibular response. Magnetic resonance imaging was normal. One year later imbalance became worse in darkness and the oscillopsia persisted. The ENG showed a left partial and a right total absence of nystagmic response to caloric (10°) and a symmetrical decreased response to pendular testing.

Evaluation of the 11 cases. In the simultaneous-BILVF group, the patients described a sudden onset of an isolated imbalance with or without acute vertigo for several days followed by a more or less permanent sensation of disequilibrium. In 2 patients, the first manifestation of BILVF was sudden falling in a dark environment, e.g. in a tunnel in one case and in the other while dancing in a dark room. One patient was exposed to a toxic substance (*case 2*), another suffered from a flu-like condition, 3 others were treated for hypertension; in one patient, the first symptoms appeared following a multiple vaccination injection (tetanus, cholera, hepatitis). In this group, except for 2 patients, all patients complained of persistent imbalance upon walking, particularly in darkness.

In the sequential-BILVF group, after an initial acute vertigo of several days' duration followed by a more or less permanent sensation of disequilibrium, patients again noted recurrent acute vertiginous episodes during the course. The first symptoms appeared following a flu-like condition in one patient treated for allergic asthma and in another patient treated for hypothyroidism (*case 4*).

In the simultaneous-BILVF group, the bilateral absence of response during caloric and pendular tests was persistent in 4 patients; 2 patients showed partial recovery with a symmetrical response to the pendular test and had a unilateral partial caloric (10°) response. The pendular test showed a symmetrical response in 2 patients without caloric recovery in one patient and with a partial bilateral caloric recovery in the other (Table II).

In the sequential-BILVF group, the ENG-findings showed partial recovery in all 3 patients: 1 patient had a symmetrical response to the pendular test with unilateral caloric recovery; in the 2 other patients, there was a bilateral hyporeflexia to pendular testing with a bilateral partial caloric recovery in 1 patient and unilateral partial caloric recovery in the other (Table III). Only 4 patients, aged from 20 to 42 years (3 in the simultaneous-BILVF bilateral and 1 in the sequential-BILVF group) complained of oscillopsia during head movements, particularly in sport activities (e.g. bicycling, skiing); this was the first symptom in one patient. Their ENG showed persistent areflexia to both pendular and caloric testing in 2 patients, a symmetrical pendular response with bilateral caloric areflexia in 1 patient and hyporeflexia to pendular testing with unilateral partial recovery in 1 patient.

Table II. *ENG-controls with a follow-up from 1 to 7 years of the patients in the simultaneous-BILVF group (n = 8/11)*

Persistent bilateral loss (pendular and caloric testing)		4
Partial recovery (symmetric response to pendular test)		4
with unilateral partial response in caloric testing	2	
with partial bilateral response in caloric testing	1	
without response in caloric testing	1	
Complete recovery		0

Table III. *ENG-controls with a follow-up from 1 to 7 years of the patients in the sequential-BILVF group (n = 3/11)*

Persistent bilateral loss (pendular and caloric testing)		0
Partial recovery		3
in caloric examination with symmetric pendular testing	1	
in caloric and pendular examinations	2	
Complete recovery		0

DISCUSSION

Oscillopsia on motion and severe imbalance in darkness are considered the two specific symptoms characterizing BILVF. It is sometimes an unexpected ENG finding in patients complaining of dizziness. BILVF is rare; it represents 0.6% of all ENG examinations performed in our laboratories. This incidence corresponds to that reported in the literature (3, 4, 6).

Regarding the clinical features and the ENG findings, we may distinguish two types of BILVF: one, with a simultaneous-bilateral vestibular loss, the other with a sequential loss of the vestibular function. In the first group with simultaneous-bilateral vestibular loss, the ENG controls performed in the follow-up showed persistent absence of caloric and pendular responses bilaterally in 5 of the 8 patients.

In the second group, after the initial acute vertigo period, patients noted several recurrent episodes of acute vertigo of variable duration and the serial ENG performed showed unilateral, then contralateral loss with or without partial recovery of function on either sides. However, except for 2 patients in the first group, all other patients experienced persistent unsteadiness worse in darkness and independent of peripheral vestibular recovery. Contrary to Baloh et al. (6), we did not observe patients with a slowly progressive form of the disease. It is possible that the progressive form postulated by Baloh corresponds to our sequential group in which we observed ipsi- and contralateral attacks in steps.

Our findings suggest that the BILVF may correspond to an acute bilateral idiopathic peripheral "vestibular neuritis" or "vertibular neuronitis" (8) occurring either simultaneously in both ears or sequentially. Schuknecht & Kimura (12) described the histopathological findings of idiopathic unilateral vestibular loss as degenerative lesions of one or more vestibular nerve branches with or without associated lesions of the end-organs. In subsequent observations, Schuknecht & Witt (5) suggested that the vestibular neuritis may also have a sequential onset of symptoms. The recovery of the vestibular function in BILVF seems to be poorer than in the case of unilateral vestibular loss (8, 10–12).

In agreement with others authors (13, 14), we observed that oscillopsia during motion was not a constant clinical complaint in patients suffering from BILVF; it represented only 36% of the cases and occurred mainly in young patients. Furthermore, oscillopsia was not strictly related to the absence of vestibular recovery as measured on ENG. Baloh (13) demonstated that in bilateral loss of peripheral sensitivity, a residual vestibular function could remain as shown by high frequency sinusoidal rotation. He also observed the rareness of oscillopsia and suggested that this was probably due to a remaining vestibular function not detectable with caloric and routine rotatory pendular examinations, which test the vestibulo-ocular reflex at low frequencies (13, 15). Others authors (14) also have reported absence of oscillopsia in patients with profound bilateral vestibular dysfunction. However, 3 of our patients had oscillopsia even with measurable nystagmic response to the rotatory pendular test at low frequency (0.05 Hz, peak velocity 60°/s). These observations suggest that the origin of oscillopsia is not completely established and that mechanisms other than the vestibulo-ocular reflex may play a role in visual stabilisation during motion (14, 16).

The etiology of the BILVF might correspond to that of unilateral sudden peripheral "vestibular neuritis" or "vestibular neuronitis". Indeed, in some cases we have noted a concomitant viral infection in the ENT area prior to the onset of BILVF. Schuknecht & Witt (5) suggested that bilateral vestibular neuritis may frequently have a viral etiology. Other factors such as exposure to toxic substances, immune diseases or reactions (17, 18), or systemic diseases such as hypertension, diabetes mellitus could also play a role in the onset of this rare bilateral vestibular pathology.

Otoneurologists should be aware of the particular clinical symptoms of BILVF which may be misdiagnosed as visual or motor dysfunction. The diagnosis is confirmed with electronystagmography and normal findings on magnetic resonance imaging.

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