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Persisting nystagmus following vestibular nerve section for Menière's disease

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Abstract
Unilateral vestibular nerve section (VNS) creates a state of acute dysequlibrium which resolves by a process of central compensation. This disturbance resolves quickly and central compensation is complete generally within a month with resolution of symptoms and signs. The course of central compensation following VNS will be similar to that seen after labyrinthectomy because the detachment of hemi-labyrinthine input that is achieved by both will be identical. Six patients are presented who have undergone VNS at least 2.7 years ago (Average 3.5 years); all of them have persisting spontaneous peripheral type horizontal jerk nystagmus, present with optic fixation in five. This is obvious clinically and was confirmed in each case by agreement of three independent observers and has been recorded by electronystagmography (ENG). They are free from marked vestibular symptoms. The explanations of mechanisms involved in central compensation are discussed with respect to this previously unrecorded clinical observation.

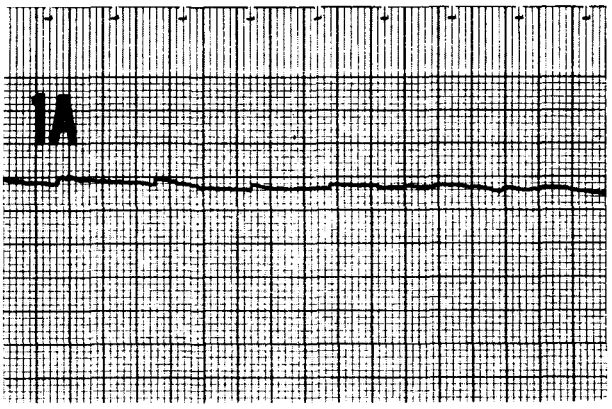
Introduction

Vestibular nerve section (VNS) has been used in the management of Menière's disease for sixty years (Dandy, 1928) and is one of the many ways of achieving anatomical detachment of one labyrinth from the central nervous system. It is indicated in cases of Menière's disease when useful hearing remains in the affected ear, and when medical management and hearing preserving surgical procedures, such as saccus decompression, have proved ineffective. It may also be indicated if, following attempted surgical destruction of a labyrinth, persisting symptoms suggest residual functioning end organ neuroepithelium, or vestibular stump neuroma (Ludman 1986). Follow-up data of patients who have undergone VNS (Barber and Ireland, 1952; Green, 1958; Kemink and Hoff, 1986) have generally concentrated upon assessment of symptomatic relief of vestibular symptoms and residual cochlear function. Only one study examined the spontaneous nystagmus, which is expected following VNS, in the immediate post-operative period (Haid, 1979), and commented upon its disappearance within two weeks of surgery.

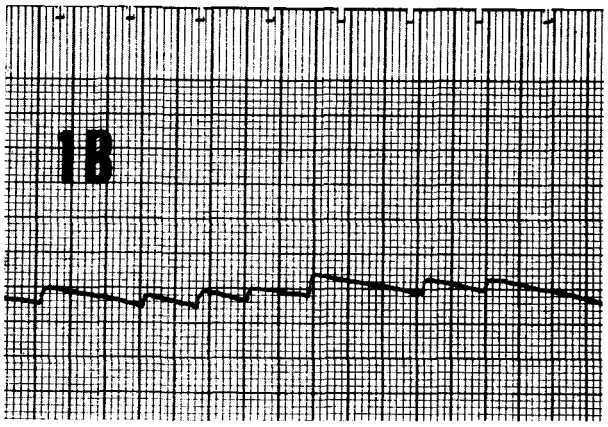
Patients

A review of six patients who had undergone VNS was undertaken. Clinically obvious horizontal jerk nystagmus was present with optic fixation (i.e. lights on), first degree to the unoperated side in five instances. This was confirmed in each case by agreement of three separate and independent clinical observers. As anticipated, this was markedly enhanced by removal of optic fixation, and became visible and recordable in the remaining patient (Case 6). ENG recordings were made in a standard fashion with an 'Ormed' machine and Direct Current amplification. The paper speed was 10 mm./second with a 10 Hertz filter and the calibration aim was for one degree of eye movement to be equivalent to 1 mm. of pen deflection. In accordance with usual convention, an upward movement of the trace indicates eye movement to the right. Representatives ENG tracings are reproduced in Figure 1. Gaze direction was limited to 30 degrees from centre. Beats of nystagmus are highlighted with arrows in two traces, 2A and 3A, where a poor high frequency response has, to some extent, diminished clarity.

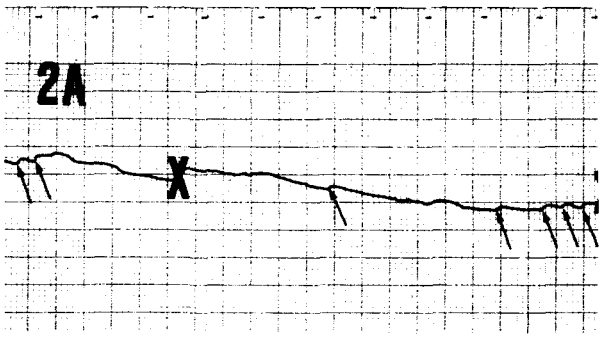
The patients' previous surgical procedures, current status and ENG findings are summarized in Tables I and II. There were five women and one man; their ages ranged from 33–66 years (average 49.9), and all had a secure diagnosis of Menière's disease made by a Consultant otologist, five had no previous otological history. The precise criteria used for the diagnosis of Menière's disease were: first, a history typical of the disorder, with each patient experiencing episodic bouts of dysequilibrium lasting between eight and twelve hours. These were associated with distortion and diminution of sound, with intolerance of loud noises; second, the demonstration of a fluctuating, yet gradually deteriorating, low tone, recruiting, sensorineural hearing loss. All patients were reviewed at least bi-annually following presentation, and for an average of 7.9 years (range 5.5–12.0) before VNS. Serial pure tone audiometry and tests for recruitment, usually a contralateral stapedial reflex at 500 Hz, were performed. A standard bi-thermal caloric test demonstrated a unilateral canal paresis in all patients prior to VNS. The remaining patient (Case 5) had a diagnosis of secondary endolymphatic hydrops, developing thirty years after a radical mastoidectomy



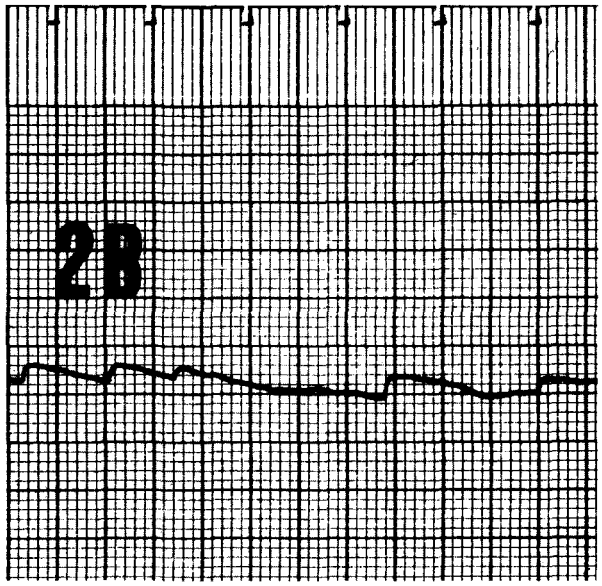
Patient 1: 1A: Looking right, with optic fixation.



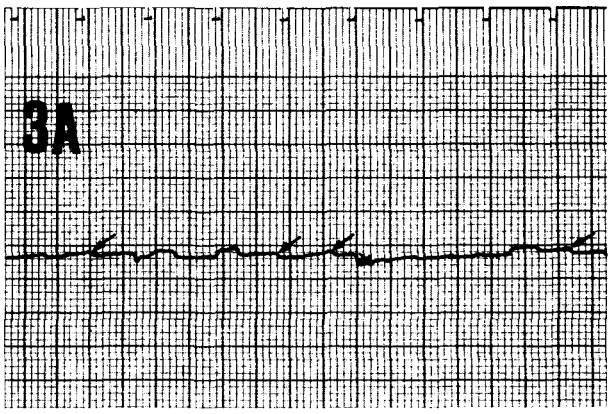
Patient 1: 1B: Looking left, without optic fixation.



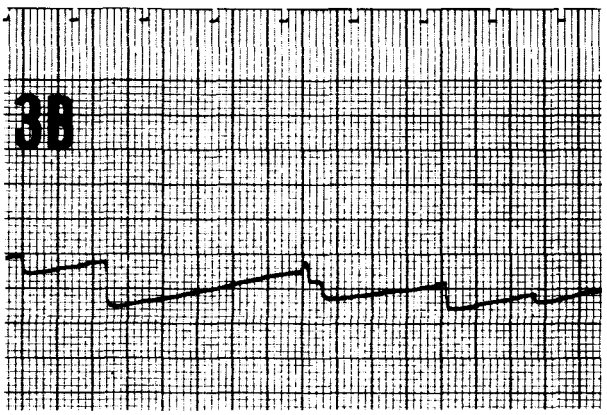
Patient 2: 2A: Looking right, with optic fixation.



Patient 2: 2B: Looking centre, without optic fixation.



Patient 3: 3A: Looking left, with optic fixation.

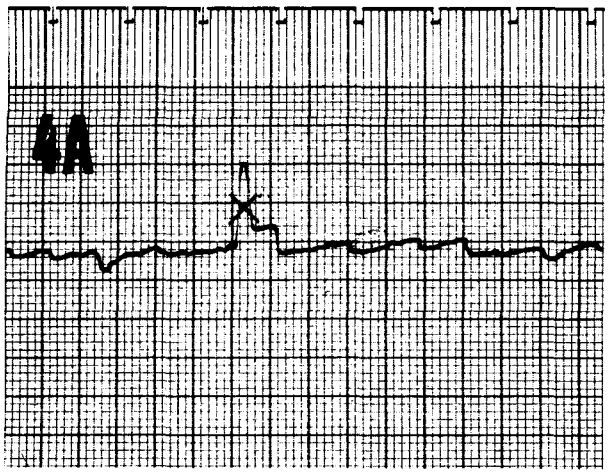


Patient 3: 3B: Looking right, without optic fixation.

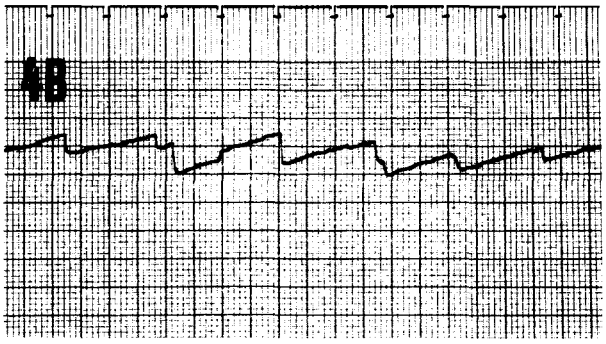
and revision procedures for cholesteatoma. Exploration of that case had excluded a lateral semicircular canal fistula which was regarded as an important differential diagnosis. A trans-mastoid 'bony' labyrinthectomy, with destruction of each semi-circular canal, had been unsuccessful in controlling the patient's symptoms.

A minimum follow-up of nine years since initial diagnosis, with normal serial pure tone audiometry and normal bithermal caloric responses in the contra-lateral ear, indicated that the Menière's disease remained unilateral in all these cases. None of the six patients had other significant diseases, and, in particular, no evidence of cerebellar pathology. Their visual fields were clinically intact, although no further, more elaborate, evaluation of visual function was undertaken.

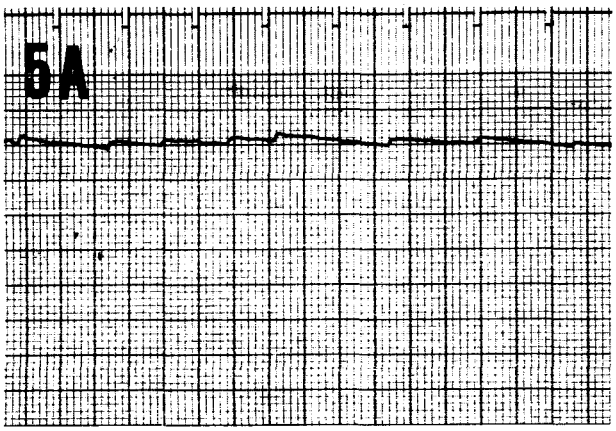
Previous surgical interventions are shown in Table I; saccus decompression was performed a total of 14 times in five cases. VNS was achieved via the suboccipital approach. The vestibular fibres of the eighth cranial nerve were identified and a segment excised at the porus



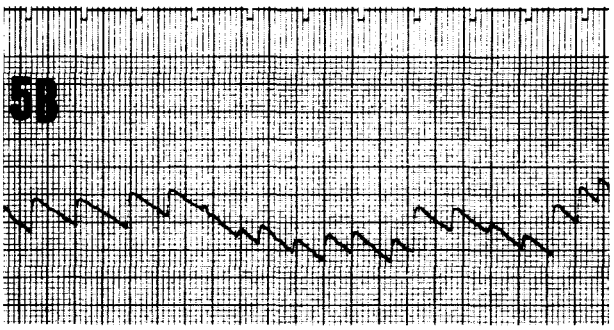
Patient 4: 4A: Looking left, with optic fixation.



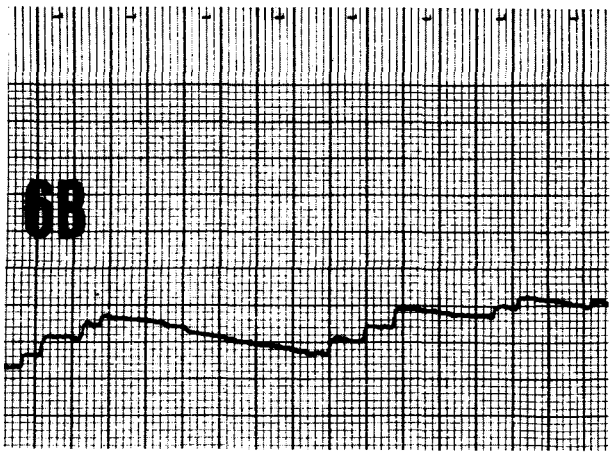
Patient 4: 4B: Looking right, without optic fixation.



Patient 5: 5A: Looking right, with optic fixation.



Patient 5: 5B: Looking left, without optic fixation.



Patient 6: 6B: Looking centre, without optic fixation.
No recordable nystagmus with optic fixation.

FIG. 1

Representative Electronystagmogram recordings.
(Note: beats of nystagmus are highlighted with arrows in traces 2A and 3A where poor high frequency response has, to some extent, diminished clarity. The crosses [×] on traces 2A, 3A and 4A indicate artifacts.)
In all cases gaze is 30 degrees left or right of centre

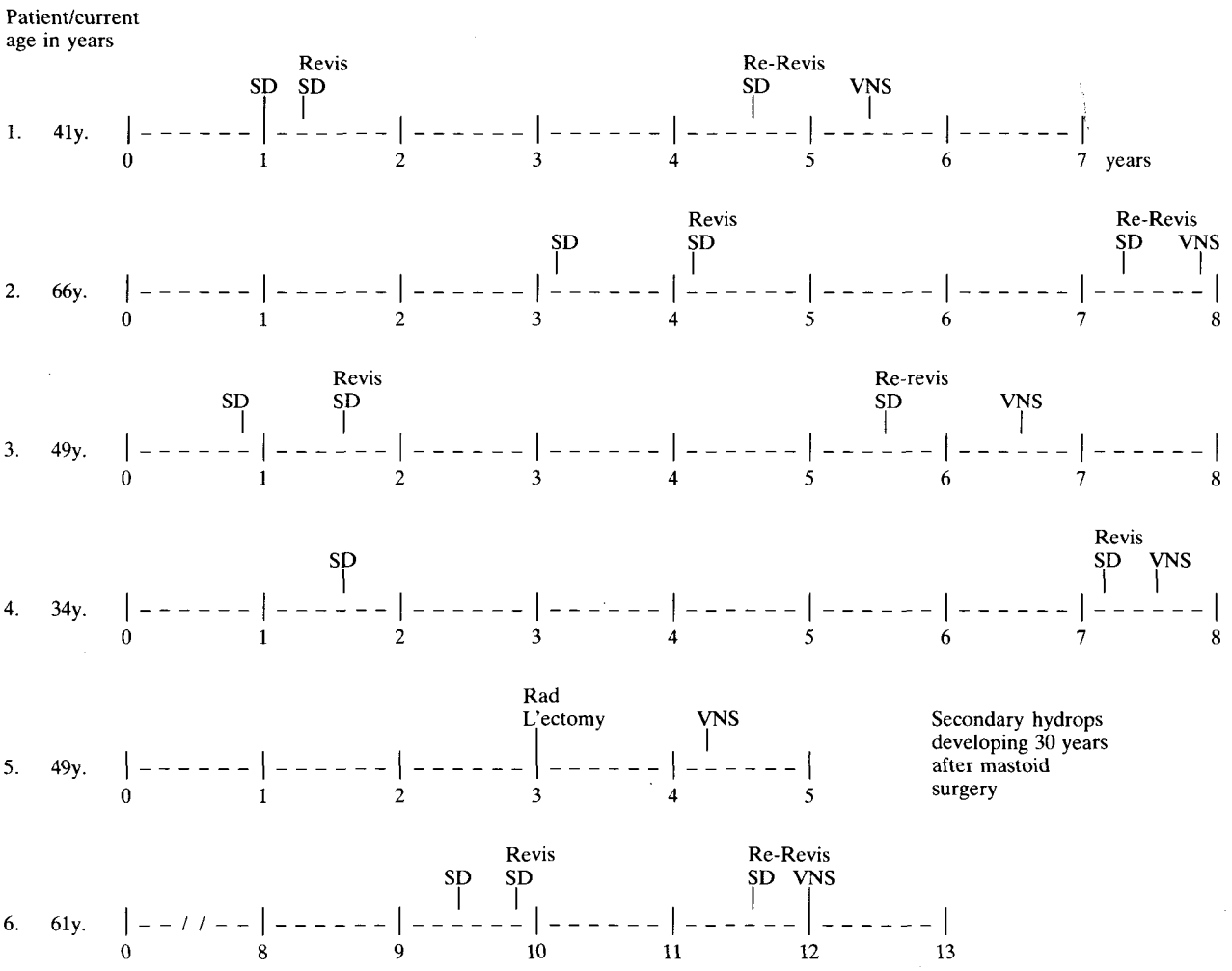
of the internal auditory meatus. In five patients, selective division of vestibular fibres was undertaken. In the remaining one (Case 5) no attempt was made to save cochlear fibres, since the hearing had been sacrificed in a transmastoid labyrinthectomy procedure.

Review of the patients took place 2.7–5.0 years (average 3.6) after VNS and four patients were seen on two occasions two months apart. These six patients were all those referred for VNS over a two and a half year period. There were no symptoms of imbalance in five patients, the sixth (Case 5) experiencing only momentary bouts of unsteadiness. Caloric stimulation with water at 20°C for

TABLE I

PREVIOUS SURGICAL INTERVENTIONS

(0 years indicates the time of initial diagnosis of Menière's disease)



Key: SD = Saccus decompression; Revis = revision; Re-Revis = Repeat revision; Rad L'ectomy = Radical labyrinthectomy.

TABLE II

NYSTAGMOGRAPHIC FINDINGS

Patient/current age in years	Side of VNS	Nystagmus		Years since VNS (years)	Persisting vestibular Symptoms
		With optic fixation	Without optic fixation		
1. 41	L	1st degree to Right	3rd degree Right	5.0	Nil
2. 66	L	1st degree to Right	2nd degree to Right	2.7	Nil
3. 49	R	1st degree to Left	3rd degree to Left	3.0	Nil
4. 33	R	1st degree to Left	3rd degree to Left	3.4	Nil
5. 49	L	1st degree to Right	3rd degree to Right	4.3	Occasional momentary unsteadiness
6. 61	L	Nil	2nd degree to Right	3.0	Nil

one minute, or Dundas Grant cold air caloric stimulation in the patient with a mastoidectomy cavity, produced no response on the operated side.

Discussion

Spontaneous horizontal jerk nystagmus persisting indefinitely after VNS in the absence of vestibular symptoms has not previously been recorded. That this is an objective observation was confirmed, in each case, by the agreement of three separate, independent, observers, and by ENG recordings. Possible explanations for the phenomenon require discussion of the mechanisms of central compensation following unilateral detach-

ment of a labyrinth from the central nervous system, however this is achieved. VNS, eighth cranial nerve section and surgical abyrinthectomy are all probably similar with regard to succeeding central events, but those following chemical labyrinthectomy, which irritates before destroying the end organ, may not be strictly comparable.

The contribution of the contralateral vestibular nuclei, the importance of alternative sensory inputs, and the effect of the cerebellum, have all been studied in various experimental animals, and the vestibulo-ocular reflex (VOR) has been examined both in humans and experimental animals.

The role of the medial vestibular nucleus, contralateral to the side of labyrinthectomy, has been studied in the cat (Precht *et al.*, 1966). Two types of neurons are described, type I, which increase their firing rate on ipsilateral rotation of the animal, and type II whose electrical activity increases on contralateral rotation. The medial vestibular nucleus contralateral to the side of labyrinthectomy assumes control of the deafferented side. This is mediated by the type II cells of the unsectioned side by a process of reduction of their inhibitory effect on the type I cells of the damaged side. The anatomical pathways involved are the internuclear commissural fibres.

The important contribution of alternative sensory inputs has been well demonstrated by depriving animals of somatosensory (Schaefer and Meyer, 1973; Lacour *et al.*, 1976) and visual input (Corjou *et al.*, 1977) when central compensation is greatly delayed.

The role of the cerebellum has been examined. Its contribution is thought to be initial inhibition of the vestibular nuclei to the extent that both sides were found to be silent following unilateral labyrinthectomy in one study (McCabe and Ryu, 1969). The term 'cerebellar clamp' has been used to describe this effect. Compensation, subsequent to the initial suppression of the vestibular nuclei, has been found to be independent of cerebellectomy (McCabe and Ryue, 1969; Haddad *et al.*, 1977), decortication, spinal cord section and attempts at reticular formation connection detachment (McCabe and Ryu, 1969). It is recognized, however, that pathways involving the cerebellum play an important part in compensation, by comparing the vestibular input with its motor effect on the external ocular muscles, and by adjusting the sensitivity of the vestibular neurons appropriately.

These are the pathways which mediate the vestibulo ocular reflex (VOR) which has also been extensively studied. This reflex maintains optic fixation when the head is moved, and is the mechanism by which eye position is maintained relative to the environment, in that any movement of the head is met with an equal and opposite movement of the eyes. It is capable of a great deal of modification. This can be shown by fitting human subjects with reversing prisms, which make the environment seem to move in the opposite direction to that anticipated. The effect upon the VOR is that, following initial suppression, it reverses its phase so that both the eye movement, and the head rotation provoking it, come to be in the same direction (Gonshor and Jones, 1976, a,b). Its pathways have been studied in the rabbit (Ito *et al.*, 1977) where it is suggested that the Purkinje cells of the cerebellum receive information both from the semicircular canals, monosynaptically, and also from the visual sensory system via the accessory olivary tract, inferior olive and climbing fibres. The Purkinje cells are said to match the electrical input from both these sources and then modulate the electrical excitability of the neurons of the vestibular nuclei so that eye position may be maintained. It might be expected that removal of input to the cerebellar Purkinje cells, either from the semicircular canals or visual sensory system, will have a profound effect upon the VOR. Complete abolition of central compensation following olivary tract destruction (Llinas and Walton, 1977) supports this suggestion, and the symptom of oscillopsia which may follow bilateral labyrinthine damage also illustrates the importance of intact VOR pathways in the maintenance of equilibrium.

Quite where this observation of persisting peripheral type nystagmus, following VNS, accords with the preceding discussion is unclear. It may be due to less than complete central compensation even though convincing vestibular symptoms are absent.

Possible explanations of this finding might include: Menière's disease in the contralateral ear, co-existent cerebellar pathology, or VOR pathway disruption.

Menière's disease in the contralateral ear is unlikely among these patients because of the normal serial pure tone audiometry and normal bithermal caloric responses in the contralateral ear in all of them over an average follow up period of 11.5 years. This comprised 7.9 years before, and 3.6 years following VNS. It is recognized, however, that these are not the only criteria that can be used in the diagnosis of Menière's disease but further, more invasive investigations, such as electrocochleography, were felt unwarranted in patients who were asymptomatic following VNS.

Significant co-existent disease, including cerebellar pathology, has similarly not been demonstrated during this period of review, and all these patients remain free from symptoms of imbalance.

The significance of VOR pathway disruption with regard to persisting spontaneous nystagmus with optic fixation following VNS requires evaluation. It is possible to theorize that, with deprivation of hemi-labyrinthine input into the cerebellar Purkinje cells, the ability of the VOR to undergo modification with regard to its phase is severely compromised, and it is the diminution of the neuronal plasticity of VOR, caused by VNS, that is responsible for the phenomenon which we describe.

Conventional teaching implies that peripheral spontaneous nystagmus of the so called 'paralytic' variety does not persist for more than a few weeks, certainly in the presence of optic fixation. The observations recorded here clearly demonstrate the inaccuracy of that belief.

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