Background, Technique, Interpretation, and Usefulness of Positional and Positioning Testing

Thomas Brandt, M.D.

TESTS FOR POSITIONAL AND POSITIONING NYSTAGMUS: AN INTRODUCTION

When the head or head and body move from one position to another, the semicircular end organ receptors and central vestibular pathways are stimulated in such a way that a compensatory eye movement occurs, helping to maintain fixation on a target during the movement. When the movement is completed, the cupulae should once again be at rest, and no imbalance in the resting afferent activity should occur. In this situation no nystagmus occurs after the head has—or head and body have—moved from point “A” to point “B.” However, if for some reason an imbalance occurs following the movement, nystagmus will result. If the nystagmus occurs as a result of the active motion of the head or head and body, the nystagmus is said to be positioning in origin. If the nystagmus occurs as a result of the new static body position, the nystagmus is said to be positional in origin. One must be aware of the precedence of spontaneous nystagmus when performing this subtest of the electrorystamnography (ENG) test battery. That is, if a patient demonstrates a coarse left-beating nystagmus on the test for spontaneous nystagmus (for example, sitting with eyes closed and alerted), it is expected that this nystagmus will contaminate the rest of the positional and positioning tests. Therefore for this patient if a left-beating nystagmus is observed during positional and positioning testing it is most likely spontaneous nystagmus and not positional/positioning nystagmus.

The positional/positioning test battery described by Barber and Stockwell included the following tests: Hallpike maneuver (head-hanging right, head-hanging left), and the following positional test positions: head and body left lateral, head and body right lateral, supine, sitting (spontaneous test), and supine with head hanging in the center position. These positions are illustrated in Fig 7–1. All of these positional test positions were conducted for at least

123
20 seconds with the patient's eyes open and then the same amount of time with the patient's eyes closed and undergoing alerting exercises in this manner, if nystagmus was noted with eyes closed but not with the eyes open, a judgment could be made as to whether the patient could visually suppress the nystagmus. A number of investigators have noted that a significant number of clinically normal individuals will generate a low-velocity nystagmus during positional testing. For instance, Barber and Wright reported that 92% (83%) of 112 healthy subjects demonstrated a position-induced nystagmus at least one of the positions with eyes closed. This nystagmus could be either direction-fixed (nystagmus beating in one direction regardless of the body position) or direction-changing (the nystagmus was right-beating when the patient was turned to the right, and left-beating when the patient was turned to the left). Given their experience, Barber and Wright empirically defined the presence of positional nystagmus as being a sign of abnormality only if (1) the direction of the nystagmus changed within a single head or head and body position, (2) the nystagmus was present and greater than 3°/sec in at least the majority of positions, or (3) if the slow-phase velocity of the nystagmus exceeded 6°/sec in a single head or head and body position. Using these criteria, the data have found that 95% of their healthy subjects would have been classified as being normal.

It is significant to note that Asch** nos described a classification system for positional/positional nystagmus. This system describes positions and positional nystagmus in terms of its duration. Type I is persistent and direction-changing. Thus, in the left lateral position the patient may demonstrate a left-beating nystagmus, and in the right lateral position the patient may demonstrate a right-beating nystagmus. Type II is direction-fixed nystagmus (nystagmus beating in the same direction regardless of the position). Both types I and II may be observed in both peripheral and central vestibular system disorders. Type III is nystagmus that stops during the time the patient is in the critical head position. The most common form of type III positional/postural nystagmus—benign paroxysmal positioning (BPPV)—is discussed in detail later in this chapter. It has been reported that it is unusual for nystagmus that accompanies BPPV to have a down-beating component, or for the nystagmus not to be paroxysmal in onset. These characteristics may point to a central vestibular system origin of this nystagmus. As discussed, with few exceptions the clinical significance of positional nystagmus is unclear. It would appear that with the exception of a nystagmus that changes direction within a single position (such as direction-changing nystagmus within a single head or head and body position), positional nystagmus that persists at equivalent slow-phase velocity with the subject's eyes closed or open (both of which are usually seen in central vestibular system disease)
that is accompanied by a nystagmus with a strong downbeat component, most types of positional nystagmus can be observed in both peripheral and central vestibular system disease. Because one purpose of conducting the ENG examination is to obtain semisubjective support for a patient's complaint, our current recommendation is to perform the positional test examination as described by Barber and Brantl (later in this chapter). If a significant positional nystagmus exists (defined by the criteria of Barber and Brantl), and the clinical importance of the finding is obscure (for example, it is not possible to differentiate the origin as being peripheral or central, or to determine whether the left or right side peripheral vestibular system is abnormal) this finding simply is presented to the referring source as semisubjective support for the patient's complaint of vertigo that occurs during a change in head or head and body position.

Chapter 7, by Dr. Thomas Brantl, reflects the changing view of the importance of positional/positioning testing. The author describes a testing protocol that is quite different from that described by Barber and Stockwell. Brantl has included only those examinations that provide information that is clinically useful. For instance, the reader is asked to note that the test protocol described by Brantl eliminates the right lateral, left lateral, supine, and head-hanging positions. The author prefers to use only three different maneuvers for both positioning and positional testing. This protocol is consistent with the modifications in the convoluted positional and positioning test that have been advocated by Barber. With eyes open and using Frenzel's lenses, the patient is moved briskly from a sitting position to a supine, head-hanging position, with the head either straight, turned left, or turned right. Brantl indicates that both positional- and position-induced nystagmus can be detected using these same maneuvers. That is, if a particular head position is maintained long enough in a single position, the positioning response ends within a few seconds and the positional response (if present) will persist for as long as the critical head position is maintained. The author reports that this method is clinically efficient in that it is not necessary to perform different maneuvers in order to elicit both positional and positioning responses.

Positional and positioning vertigo and nystagmus syndromes can be attributed to either peripheral or central vestibular dysfunction (Table 7–1). Positional and positioning testing are an integral part of the examination of balance function even if the patients do not complain about symptoms associated with head motion. Simple observation of positional and positioning nystagmus by use of Frenzel's lenses often allows one to establish the diagnosis. ENG recording of the eye movements evoked by these tests provides reliable documentation but does not contribute substantially to the success rate of diagnosis.

POSITIONAL AND POSITIONING TESTING

Careful history-taking is the key to diagnosis and is still much more important than electro-oculography or modern brain imaging techniques, in the evaluation of vestibular system disease. The examiner should attempt to elicit from the patient a report of head motion intolerance or of vertigo (for example, positioning vertigo), oscillopsia, and nausea associated with changes in head position (for example, positional vertigo).

Positional/Positioning Test Technique

A distinction should be made between a positioning response and a positional response. Positioning nystagmus

*Barker HD. Positional nystagmus. ENG Re

and vertigo are precipitated by rapid head extension or lateral head tilt which evokes an enhanced postrotatory re- sponse. This may be contrasted with po- stional nystagmus and vertigo, which are due to the position of the head rela- tive to the plane of gravity rather than the velocity of head movement.

Positioning and positional testing may be detected with the same maneu- vers. The test protocol is summarized in Table 7–2. The most provocative posi- tioning test is to briskly tilt the patient as a unit from a sitting to a supine, head-hanging position with the head straight or turned to the left or to the right. An alternative procedure is to tilt the sitting patient laterally by grasping the head bitemporally (Fig 7–2). If the examiner observes the eye movements long enough following the head tilt, then he or she will be able to recognize both positioning responses (which cease after a period of time that may last from a few seconds to a maximum of 1 minute) and positional responses (which persist as long as the head position is maintained). Eye movements should be observed in each position for at least 20 seconds (if no nystagmus occurs) or up to several minutes (if nystagmus oc- curs). In this way, it is possible to differ- entiate between paroxysmal positioning nystagmus and central vestibular posi- tional nystagmus (positional nystagmus that does not attenuate with visual fixa- tion). Positioning maneuvers, if provoca- tive of nystagmus or vertigo, are re- peated several times in both directions (right ear down, upright, left ear down) in order to evaluate carefully the posi- tion-induced response in terms of la- tency, duration, reversal, and fatigabi- lity of the nystagmus. Eye movements (in primary position and or lateral gaze) are observed both in the offending head

Table 7–1.

Vertigo and/or Nystagmus Associated With Head Motion or Changes in Head Position Relative to the Gravitational Vector

<table>
<thead>
<tr>
<th>Positional vertigo</th>
<th>Central vertigo</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vertigo and/or nystagmus</td>
<td>Vestibular system</td>
</tr>
<tr>
<td>Central position</td>
<td>Positional nystagmus</td>
</tr>
<tr>
<td>Upright nystagmus</td>
<td>Central positional nystagmus without vertigo</td>
</tr>
<tr>
<td>Initial positional vertigo with nystagmus</td>
<td>Basilar insufficiency</td>
</tr>
<tr>
<td>Vertigo due to nystagmus compression or “disabling positional vertigo”</td>
<td>Vestibular nevus</td>
</tr>
<tr>
<td>Benign paroxysmal positional vertigo</td>
<td>Cupulolithiasis and/or heavy head</td>
</tr>
<tr>
<td>Positional vertigo</td>
<td>Positional head extension vertigo or bending over vertigo</td>
</tr>
<tr>
<td>Positional nystagmus</td>
<td>Vestibular head movement intolerance (dizziness and unsteadiness of gait)</td>
</tr>
<tr>
<td>Positional nystagmus with macroglossus (tongue)</td>
<td>Bilateral vestibulopathy</td>
</tr>
<tr>
<td>Benign paroxysmal positional vertigo</td>
<td>Ocular motor disorders (defective vestibulo-ocular reflex)</td>
</tr>
<tr>
<td>Positional nystagmus with macroglossus (tongue)</td>
<td>Neurovascular cross-compression (vestibular paroxysmy)</td>
</tr>
<tr>
<td>Benign paroxysmal positional vertigo</td>
<td>Vestibuloocular reflexes</td>
</tr>
<tr>
<td>Vestibuloocular reflexes</td>
<td>Perilymphatic fistula</td>
</tr>
<tr>
<td>Post-traumatic otolith vertigo</td>
<td>Vestibulocochlear injury (e.g., alcohol, phenytoin)</td>
</tr>
</tbody>
</table>
position and after return of the patient into the Primary sitting position.

**Spontaneous Nystagmus**

The spontaneous nystagmus test protocol is shown in Table 7–3. The patient’s eyes are first inspected with his head stationary at a normal upright position and with fixation on a target straight ahead. Spontaneous vestibular nystagmus and other ocular oscillations that are present when the patient is in the static, sitting position are often confounding to clinicians because these results contaminate positional and positioning testing. Spontaneous vestibular nystagmus of peripheral origin is exaggerated or becomes overt when fixation is removed. Therefore, with the patient in
the same static position, Frenzel’s lenses are used for evaluation of involuntary ocular oscillations in the absence of fixation. Evaluation of the positional response may be difficult in all patients suffering from additional spontaneous nystagmus or other ocular oscillations that override fixation. Normal vestibular end organs generate testing firing frequency that is the same bilaterally. This continuous excitation is transmitted to the vestibular nuclear by way of vestibular nerves. Pathologic processes affecting an end organ or the vestibular nerve alter its firing frequency, thereby creating a tone imbalance. The imbalance causes nystagmus and vertigo. Therefore, spontaneous nystagmus denotes the presence of a tonic imbalance in the peripheral vestibular system.

Head-Shaking Nystagmus

(High-Frequency Testing)

The well-known clinical method of provoking spontaneous nystagmus by passive head shaking of a patient wearing Frenzel’s lenses reveals a unilat- eral labyrinthine loss even if it is apparen- tly centrally compensated. The patient is asked to vigorously shake his or her head side-to-side (horizontal gaze plane) or vertically (pitch plane) for 10 seconds before the eyes are observed for transient nystagmus. Hain et al.28 were able to show that horizontal head shaking in the yoke plane elicits horizontal nystagmus with slow phases that are initially directed toward the side of the lesion and upward. These patient’s eyes are examined to determine the horizontal and vertical vestibular function. The head is rotated rapidly toward the side of the lesion, patients with unilateral vestibular loss make clinically evident, as shown in Fig. 7-3. of Halmagyi and Curthoys10 observed that there is no central compensation of the positional asymmetry (for example, unilateral abnormality) of high-frequency canal function. When the head is rotated rapidly toward the side of the lesion, patients with unilateral vestibular loss make clinically evident, opposite directed, compensatory nystagmus. This indicates a unilateral high-frequency defi- ciency of the VOR, produced by function of the remaining labyrinth. This simple bedside test is also useful for the detection of bilateral vestibular loss.

If the head is passively turned quickly (say faster than 1 Hz and 100°/s peak acceleration) then the compensatory eye movement to maintain gaze in space is overriding the vestibular system rather than the optokinetic pursuit reflex or vestibulo-ocular reflex. With bilateral vestibulopathy, despite attempted fixation on a stationary target, the gaze shifts with the head. Because compensa- tory eye movements are inappropriate, gaze in space is corrected after the head movement by a compensatory saccade toward the fixation target (see Fig. 7-3), which can be easily detected by the observer. These tests do not exclude the possibility that parts of the vestibular
labyrinth may still function, especially the vertical semicircular canals or the otoliths.

**CLINICAL ENTITIES: POSITIONING TESTING**

**Benign Paroxysmal Positioning Vertigo**

In benign paroxysmal positioning vertigo (BPPV), initially described by Bárány in 1921, brief attacks of rotary positional vertigo and concomitant positioning rotatory-linear nystagmus are precipitated by rapid head extension as well as by lateral head tilt toward the affected ear. It is the most common cause of vertigo in the elderly and is due to cupulolithiasis of the posterior semicircular canal [14, 116] or by canalolithiasis.

In these patients there is a frequent history of spontaneous recovery, and, there exists the possibility of a most effective mechanical therapy by performing positioning maneuvers. The diagnostic criteria for BPPV (Table 7–4) are based on the time history of the burst of rotational vertigo and sometimes nausea associated with the typical positioning nystagmus. Both symptoms are induced by rapid head and body movements from the sitting to the head-hanging right or left position. Other criteria include:

1. Latency. Vertigo and nystagmus begin 1 or more seconds after the head is tilted toward the affected ear, and an increase in severity to a maximum.
2. Duration less than 1 minute. Nystagmus reduces gradually after 10 to 40
TABLe 7—4. Diagnostic Criteria for Benign Paroxysmal Positioning Vertigo

Clinical syndrome
Brief attack of rotatory vertigo and rotary-linear nystagmus precipitated by rapid head turn toward the affected ear, or by head extension. Typical rotary nystagmus that beats toward the uninvolved ear, has a latency of 1 to 3 sec, a duration less than 1 min, a reversal on righting, and fatigues when prolonged.

Incidence/age/gender
Direction of fall is toward the affected ear and forward. Idiopathic forms (50%): Most common cause of vertigo in the elderly (6th and 7th decades); female: male ratio = 2:1. Symptomatic forms (50%): e.g., post-traumatic, postural or neurologic (cerebellar, stroke or aneurysm), female: male ratio = 1:1.

Pathomechanism
"Cupulolithiasis or caloricolithiasis": dislodged otolithia (degeneration, trauma) rests in the posterior semicircular canal and cause gravity dependent cupula deflection.

Course/prognosis
Mostly benign with spontaneous recovery within weeks or months (70%), but persistent (without treatment) in about 20% to 30%, or shows recurrence at variable periods for years.

Management
Most effective physical therapy by positioning maneuvers on a serial basis; in rare intractable cases, plugging of the posterior semicircular canal.

Differential diagnosis
Central postmict nystagmus/vertigo, perilymphatic fistula, drug or alcohol intoxication, Meneire's disease, neurovascular crisis, compression, Waldeyer's disease, psychogenic vertigo.

seconds and ultimately abate even with maintenance of the precipitating head position.

3. Linear-rotatory nystagmus. The nystagmus is best seen when the patient is wearing Frenzel's lenses (for example, 20 dioptries lenses), which prevent suppression by fixation. The cupula is linear-rotatory, with the fast phase beating toward the uninvolved ear or upward when the patient's gaze is directed toward the uppermost ear (Fig. 7—4).

4. Reversal. When the patient returns to the seated position, the vertigo and the nystagmus may recur less violently in the opposite direction.

5. Fatigability. Constant repetition of this maneuver will result in ever lessening symptoms.

During the initial course of the disease, when patients are standing in the normal head-upright position they may complain of other symptoms of otolithic vertigo. These include the sensation of feeling as if they are walking on pillows. This sensation probably arises from the unequal excess loading of the two utricular otoliths. Central mechanisms provide compensation within one to three weeks.16,17

Mechanisms Underlying BPPV: CupulolithiAsis
The early assumption by Barany116 and Dix and Halpike26 that the underlying lesion must be situated in the vestibular end organ and must involve the otoliths was later supported by Schuknecht113,115 and Schuknecht and Ruby.116 Schuknecht and his co-workers postulated a mechanical pathogenesis termed "cupulolithiAsis."4 They found basilar deposits on the cupula in the posterior semicircular canals to patients who manifested unilateral BPPV prior to death from unrelated disease. These deposits exceeded the size of those found in more than 30% of temporal bones in a control population. They argued that inorganic particles, detached from the otocutal layer by spontaneous degeneration or head trauma, gravitate to and become settled on the cupula of the posterior semicircular canal, which is situated directly inferior to the utricle when
the head is upright (Fig 7–5). The posterior semicircular canal thus serves as a receptor for the detached sediment. In fact, otoconia are easily dislodged by linear accelerations or centrifuging in animals.10,12 Otoconial debris become displaced in old age,73 and lodge either in the posterior semicircular canals128 or in the cochlea in cochleosaccular degeneration.128

The cupula normally has the same specific gravity as the endolymph and is a transducer of angular accelerations only. When heavily loaded, it should theoretically become sensitive to changes in head position relative to the vertically oriented earth gravitational field (gravitational vector; see “the buoyancy hypothesis” later in this chapter). The common view is that BPPV simply reflects transformation of the affected cupula from a transducer of angular acceleration to one of linear acceleration, secondary to the acquired specific gravity differential (“heavy otoconial debris acting on cupula”) between the cupula and endolymph.103,113 This view cannot be correct for several reasons: BPPV is a positioning rather than a positional vertigo/ nystagmus because it is induced only by rapid changes in head position, and the paroxysmal nystagmus is compatible with the cupulogram of an ampullofugal stimulation of the posterior semicircular canal rather than with ampullopetal positional effects as expected. Schmidt112 was able to reverse the direction of paroxysmal positioning nystagmus by making his patients bend rapidly forward. On the bases of utriculo-cupula interaction as described by Flum,106 he argued that BPPV is the consequence of a disinhibited angular reaction of the posterior canal arising from disturbed utricular function. We believe, however, that either cupulolithiasis or canalolithiasis are
Fig 7-1. Mechanism of cupulolithiasis, as a possible cause of benign paroxysmal positioning vertigo. Inorganic "heavy" particles detached from the ocular layer (by degeneration or head trauma) gravitate to and become settled on the cupula of posterior semicircular canal. The "heavy" material causes a specific gravity differential between cupula and endolymph with postrotatory overexcitability. After rapid head tilt towards the affected ear or after head extension, when the posterior semicircular canal is moved in the specific plane of stimulation, an ampullopapillary deflection of the cupula occurs with rotational vertigo and concomitant nystagmus.

Causative, in that the "heavy cupula" creates an overexcitability of the posterior canal to angular accelerations 1, 2, or a free-floating clot of inorganic particles (heavier than endolymph) gravitates and produces push or pull forces on the cupula.

The intensity of positional nystagmus depends on the velocity of the positioning maneuver, and BPPV attacks can be avoided if the challenging position is assumed very slowly (longer than 6 seconds).

The suggestion that the posterior semicircular canal plays the critical role in BPPV is in accordance with the finding that an ampullopapillary cupula deflection causes a contraction of the ipsilateral superior oblique and the contralateral inferior rectus eye muscles. 12, 13 It is also consistent with the pattern of eye movements produced by selective stimulation of the posterior semicircular canal, as demonstrated in the monkey by Cohen et al. 32 and in man by Morgenstern and Forrenkopf. 36 It is further supported by Gacek, 37 who cured patients with chronic unilateral BPPV by selective dissection of the ipsilateral posterior ampullary nerve. The presence of cupulolithiasis and endololithiasis has been shown histologically, and it could provide an explanation of tinnitus. The detached particles could disperse from the cupula into the endolymphatic space, and this would account for the diminution of the symptoms.

Etiologic Condition
In the early stages, BPPV is usually experienced on awakening in the morning rather than on first lying down. Studies in large series of patients 11, 80 support the common clinical finding that the following conditions figure in the etiology of BPPV: head (labyrinthine) trauma, viral neurotubulinrithis, vertebrobasilar ischemia (anterior inferior cerebellar artery), post-surgery (ear and general), prolonged bedrest due to unrelated diseases, and most often idiopathic causes (aging).

In the series of 240 patients described by Baloh et al., 11 the origin of BPPV was idiopathic in about half of the cases. In the remainder the most commonly identified causes were head trauma (17%) and viral neurotubulinrithis (15%). We found that out of a total of 104 patients with unilateral BPPV, 12% had suffered from vestibular neuritis days to years previously. 7 The age at onset of BPPV ranges from adolescence to old age; and in the idiopathic group exhibits a peak incidence in the 6th and 7th decades. In contrast, onset tends to be earlier on av-
erage in symptomatic forms of BPPV. There is a striking female preponderance exceeding a ratio of two women to one man in the idiopathic group, whereas sexes are about equally distributed in the post-traumatic and postviral neurological forms.

When patients present with post-traumatic BPPV, it is sometimes difficult to determine retrospectively whether the trauma really caused the vertigo or vice versa. In about 10% of the cases, observed BPPV is bilateral (mostly asymmetrical) and is particularly associated with post-traumatic forms. The relatively frequent coincidence of BPPV following vestibular neuritis was first explained on the basis of ischemia of the anterior vestibular artery. However, it is more likely to be due to viral inflammation of the vestibular nerve, and in some cases promotes the concept that vestibular neuritis results in a partial rather than complete unilateral vestibular paresis. This may occur because BPPV requires preserved function of the posterior canal. The natural history of BPPV is considered benign because of its spontaneous recovery within weeks or months, but in about 20% to 30% of the patients the condition persists (when untreated) or reoccurs after variable periods for years.

**Nystagmus**

Visually observed paroxysmal positioning nystagmus (best seen with the eyes behind Frenzel's lenses) is similar in direction in all cases, and is beating toward the undermost and affected ear, with a rotatory component clockwise (when following leftward movement) or counterclockwise (when following rightward movement), as seen by the investigator. As indicated earlier, these patterns of eye movements and the characteristics of a short latency, limited duration, reversal on return of the patient to the upright position, and fatigability on repetitive provocation are sufficient to establish the diagnosis.

A closer look however, in particular at the gaze-dependent differential effects on the direction and the conjugation of induced eye movements, reveals much more complexity and explains some of the seemingly contradictory descriptions in the literature. What Herbert rediscovered in 1970 was already part of the original description by Barany in 1921. This was that positioning nystagmus is mainly rotatory and beats toward the undermost ear when gaze is directed toward the undermost ear, but beats mainly in a linear-oblique direction toward the forehead when the gaze is directed toward the uppermost ear.

The neuronal network mediating the VOR of the vertical canals is based on sensory convergence within a three-neuron reflex arc. It links a set of extracocular muscles with their primary action aligned to the particular spatial plane of either the anterior or posterior canal. Sensorimotor transformation from canal planes to the plates of eye movements has been demonstrated at the level of second-order neurons within the vestibular nuclei. These neurons projecting to oculomotor neuron pools always contact their two respective principal eye muscles in the excitoria as well as in the inhibitory (push-pull operational mode) vestibulo-ocular motor link.

Ampullolugal stimulation of the posterior semicircular canal causes excitation of the ipsilateral superior oblique and the contralateral inferior rectus muscles. This causes both eyes to move downward with the slow phases and to move upward with the quick phases of the nystagmus (see Fig. 7-4).

Monocular ENG recordings of horizontal and vertical components demonstrated a larger horizontal component in the ipsilateral eye and a larger vertical component in the contralateral eye, both of which can be explained by the different angle of insertion of the oblique and rectus muscles. Furthermore, the amplitude of the horizontal component in each eye depends on the direction of gaze relative to the head.
and ENG recordings sometimes appear inconsistent with visual observation, such that reversals of the horizontal component with changes in gaze position and dissociated eye movements as well as downbeating nystagmus were reported.11

Vertigo and Postural Imbalance

The occurrence of BPPV with the patient supine is very discomforting and makes patients afraid of falling backward, an almost unique complaint. In the upright body position vertiginous attacks produced by changes in head position are incapacitating and can be dangerous, such as when a patient is looking up at the ceiling while standing on a ladder. In this situation, BPPV can cause a catastrophic fall. Although the nystagmus pattern in BPPV attacks has been well described, less information is available on the direction and magnitude of postural decompensation.

Posturographic measurements have been made in patients with BPPV in whom attacks were elicited by head tilt while they were standing on a posturographic platform. These measurements revealed a characteristic pattern of postural instabilitya: after a short latency, patients exhibit large sway amplitudes, predominantly forward-and-backward (Fig 7–6), with a mean sway frequency range below 3 Hz. The amount of instability decreases gradually within 30 seconds in parallel to the diminution of the sensation of vertigo and of the nystagmus. When subjects close their eyes, for tendency to fall can hardly be compensated for by interference of corrective somatosensory input. Patterns standing on a force-measuring platform show a shift of the mean position of the center of gravity forward and toward the direction of the head tilt (see Fig 7–6), with a concurrent increase in sway amplitude. Mean sway amplitudes increase by a factor of 4 in the forward-backward plane and a factor of 3 in the lateral plane. Some patients exhibit a superimposed 3 Hz forward-backward sway, an oscillator-like tremor of body posture as described in chronic alcoholics with cerebellar atrophy.19

The measurable shift of the center of gravity in the forward direction, and ipsilateral to the tilted head, can be interpreted as the motor compensation for the initial subjective vertigo in the opposite direction, the diagonal plane being identical to the spatial plane and working range of the ipsilateral posterior canal.20 Thus, compensation for the initial perceived (subjective) fall caused by the lesion occurs in such a way that a measurable (objective) fall results in the opposite direction. Posturographic data are consistent with the hypothesis that the posterior canal is responsible for the
generation of paroxysmal peripheral position vertigo.

Posturographic measurements performed by Black and Nashner\(^7\) were unable to distinguish three groups of patients: those with unilateral or bilateral loss of peripheral vestibular function, those with BPPV without peripheral vestibular deficit, and those with the combination of both BPPV and peripheral vestibular deficit. Patients were exposed to either a stable or a moving foot support, and to variable visual surroundings. The authors concluded from their results that the BPPV patients principally use visual information to compensate for the postural destabilization induced by a vestibular irritation, whereas a peripheral vestibular deficit causes a disturbed adaptive reorganization of visual and postural references for orientation in space. Vestibulospinal compensation results, therefore, from either suppression of vestibular inputs or from simultaneous selection of an alternative reference for orientating vision or support. Clinicians are well aware that patients in the acute phase of BPPV also complain of unsteadiness in gait and postural balance, which they describe as "walking on pillows." These symptoms can be classified as otolithic vertigo, and can be attributed to the suddenly unequal weights on the maculae of the two utricles which generate a vestibular tone imbalance. Impairment of control of posture and gait usually shows a gradual improvement in the following days or weeks, which reflects either central compensation or peripheral restitution of the equal weights. It is not easy to examine utricular function in isolation in human subjects, but one possible source of information is dynamic ocular counterrolling, a reflex that appears to depend on the utricular otolith organs. Mushinski et al.\(^8\) described abnormal ocular counterrolling in 16 of 18 patients with BPPV tested by rotating the body about two axes with the head fixed in relation to the body. The two axes were the nasa-occipital and the submental-vertex (barbecue rotation), which provided a more sensitive indicator of utricular dysfunction than the nasa-occipital. The most common dysfunctions were disconjugate eye movements and hypometricity. Further, subjects were more sensitive to ipsilateral tilt than to contralateral. This agrees with our own experience in patients with acute BPPV, who show significant deviations of the subjective visual vertical when they adjust a test bar to perceived vertical.

Management

Based on the mechanical hypothesis of cupulaithus we constructed an effective physical therapy for these patients in order to promote the loosening and ultimate dispersion of the degenerated otolith material from the cupula of the posterior canal.\(^6\) Specifically, patients are instructed to provoke systematically vertigo attacks by repeatedly tilting their upper trunk and head in the challenging position, with the lateral aspects of their occiput resting on the bed. This ensures proper plane-specific stimulation of the posterior semicircular canal. They remain in this position until the evoked vertigo subsides, or for at least 30 seconds and then by a 180° assumption the opposite head-down position for a further 30 seconds. The sequence of positioning is repeated about five times during each session. The maneuvers are carried out by the patients themselves every 3 hours while awake, and are terminated after 2 consecutive vertigo-free days. In extreme cases, when patients are subject to nausea, or in particularly anxious patients, vestibular sedative drugs such as dimenhydrinate or scopolamine are given during the first 1 to 3 days of the physical therapy.

This simple physical approach has lead to relief in the majority of cases\(^6\) within 1 to 4 weeks (Fig. 7-7), even if the vertigo had lasted for months before the initiation of the therapy. In post-traumatic forms a slight but minimally distressing BPPV may persist
which is unresponsive to physical training. The time course of individual recovery, undulating and with abrupt remissions, supports a purely physical mechanism rather than central compensation by habituation (see Fig 7–7).

In the rare patients not responding even to prolonged physical therapy, surgical transection of the posterior ampullary nerve by a middle ear approach can be considered.105,106 This operation provides relief of vertigo; it is, however, not easy to locate the posterior ampullary nerve surgically, and sensorineural hearing loss is one of several possible postoperative complications.107,108 In our experience with hundreds of BPPV patients there are only a few individuals who did not respond well to physical therapy and who ultimately required selective surgical transection of the posterior ampullary nerve. Recently, an alternative surgical procedure of transmastoid posterior semicircular canal occlusion has been described for intractable BPPV.109 Drug therapy with anti-motion sickness medications reduces nausea but has not been proved to be a particularly efficacious treatment of BPPV.

**Horizontal Semicircular Canal BPPV?**

McClure106 described paroxysmal nystagmus in some patients which possibly originated from stimulation of the horizontal semicircular canal. This was supported by Katsarkas,74 who reported a small number of patients with unusual paroxysmal positioning nystagmus (for
example, it may be purely horizontal or rotary-linear, with the fast phases in the opposite direction to that usually observed. This is probably compatible with excitation of the posterior or horizontal semicircular canal of the lowermost ear, or with excitation of the superso- rior canal of the uppermost ear. In four of five patients with suspected horizontal canal BPPV simular rotational tests revealed a reduced phase shift of eye velocity relative to head velocity at 0.04 Hz compared with normal subjects. This change was attributed to either reduced elasticity of the cupula or increased viscoelastic friction of the canal fluid.

**Bending-Over Vertigo**

A combination of mechanisms similar to that described for head-extension vertigo (discussed later in this chapter) would be applicable in explaining the common symptom of vertigo upon bending over at the waist. An added consideration is the transient increase in intracranial pressure (secondary to increased cephalic venous pressure) being transmitted to the perilymphatic space surrounding the endolymphatic membrane.

**Clinical Entities: Positional Testing**

**Spontaneous Nystagmus: Peripheral Vestibular System Disease**

Spontaneous vestibular nystagmus is mostly horizontal-rotatory (clockwise, left beating; or counterclockwise, right beating). The nystagmus is typically reduced in amplitude by fixation (fixation suppression) and enhanced by eye closure or use of Frenzel’s (high plus di- opter) lenses. According to Alexander’s law, spontaneous nystagmus is increased with gaze shifts toward the fast phase, and decreased with gaze shifts toward the slow phase of the nystagmus. This may mimic gaze-evoked nystagmus in a patient with moderate spontaneous nystagmus which is completely suppressed by fixation straight ahead but still present with the gaze directed toward the fast phase. Positional testing may unspcifically facilitate nystagmus.

In the acute stage of peripheral labyrinthine vertigo, vestibular system disease (for example, vestibular neuritis, herpes zoster oticus), horizontal-rotatory spontaneous nystagmus beats toward the unaffected ear. After recovery, spontaneous nystagmus in some patients transiently reverses its direction (Ehrliehnynystagmus or recovery nystagmus) when the centrally compensated lesion regains function. The direction of spontaneous nystagmus is less conclusive in other labyrinthine disorders such as Meniere’s disease or perilymph fistulas.

**Spontaneous Nystagmus Central Nervous System Disease**

A spontaneous nystagmus that is purely horizontal (for example, as opposed to horizontal rotatory) in direction and not attenuated by fixation is caused by a central lesion. More often central spontaneous nystagmus is mixed linear-rotatory or purely rotatory in vertical. It is evoked by acute lesions adjacent to the vestibular nuclei, such as in posterior medullary infarction (Wallenberg’s syndrome).

**Periodic Alternating Nystagmus**

This occurs with a variety of cerebellar conditions and is a spontaneous horizontal nystagmus that reverses its direction approximately every 2 minutes.

**Acquired Pendular Nystagmus**

This type of nystagmus is usually a small-amplitude, vertical (torsional) oscillation at a frequency of 2 to 7 Hz, sometimes disconjugate or purely monoocular. Multiple sclerosis is the most common cause.
Downbeat Nystagmus

When downbeat nystagmus occurs in the primary position of gaze, or more particularly on lateral gaze, it is often accompanied by oscillopsia and postural instability. It is due to a lesonal tonic imbalance of the vertical VOR in the pitch plane and is almost specific to structural lesions of the paramedian craniovertical junction (25% of patients with Arnold-Chiari malformation).

Upbeat Nystagmus

This type of nystagmus in the primary position of gaze with concordant oscillopsia and postural instability is a pendular of downbeat nystagmus and most probably reflects an imbalance of vertical VOR in the pitch plane. Static head tilt to the prone and supine positions modifies the characteristics of upbeat nystagmus in most cases. It may be enhanced but is mostly suppressed (Fig 7–8). Two separate brain stem lesions in the pontomesencephalic junction and in the medulla near the perihypoglossal nuclei are likely to be responsible for this syndrome.

Congenital Nystagmus

Congenital nystagmus is usually horizontal, with abnormal wave forms. It is activated by fixation, modulated by gaze direction, and diminished by convergence. Some patients exhibit additional head oscillations or a head turn; changes in head position relative to the gravitational field are ineffective. An inverted optokinetic nystagmus (as tested by an hand-held rotatory optokinetic drum) is almost pathognomonic. History taking is most important, as the condition is usually diagnosed during infancy.

Spontaneous Nystagmus: Influence of Drugs/Medications

The list of drugs that may have adverse effects on eye movements and balance is impressive (Tables 7–5 and 7–6). The administration of these drugs in therapeutic doses may result in spontaneous nystagmus.

Positional Nystagmus Vertigo

With Specific Gravity Differential Between Cupula and Endolymph: The Buoyancy Hypothesis

Transient positional nystagmus has been repeatedly described following the ingestion of water-soluble molecules with differing specific gravities, such as alcohol or heavy water. The semicircu-
lar canals selectively transduce angular velocity and acceleration, and under normal circumstances are insensitive to gravitational orientation and linear acceleration. A major reason for the lack of sensitivity is that the cupula and endolymph have the same specific gravity (the sensory hair cells are embedded in the cupula, which is housed in the ampulla of the canals). The neutral buoyancy of the cupula in the endolymph prevents any out-of-balance forces when linear accelerations are applied. If a considerable specific gravity differential occurs between cupula and endolymph, then the semicircular canals should become sensitive to changes in head position within the gravitational field (see Fig. 7–9), resulting in positional rotatory vertigo and nystagmus. The direction of nystagmus and vertigo should be dependent on the particular head position (according to the different planes of the horizontal and vertical semicircular canals) and on whether the specific gravity of the cupula is greater or less than that of the endolymph. Thus, nystagmus should be direction-changing with either head position right lateral or left lateral, beating toward the undermost ear, with the cupula heavier than endolymph.

This hypothesis, known as the buoyancy hypothesis, requires that ingested compounds of different specific gravity diffuse at different speeds into the cupula and endolymph, thus causing the transient density gradient. Experiments were prompted with ethanol,211 diuretic oxirane,212 and glycerol,197 which all induced positional nystagmus consistent with the hypothesis.

Several questions are raised when considering the pathophysiological concept of a gravity differential between cupula and endolymph which causes either a positional (cig (cig), glycerol, heavy water, macroglobalinemia) or a positioning vertigo (nystagmus (cupulolithiasis)). Why is positional alcohol nystagmus (see Fig. 7–9) maintained over minutes and hours in the precipi-
<table>
<thead>
<tr>
<th>Drug and toxic agents</th>
<th>Oculomotor abnormalities</th>
</tr>
</thead>
<tbody>
<tr>
<td>Phenylm</td>
<td>Gaze-evoked nystagmus, saccadic pursuit, external ophthalmoplegia, internuclear ophthalmoplegia. Rare: periodic alternating nystagmus, downbeat nystagmus, vertigo.</td>
</tr>
<tr>
<td>Carbamazepine</td>
<td>Gaze-evoked nystagmus, saccadic pursuit, impairment of optokinetic nystagmus, external ophthalmoplegia, downbeat nystagmus, vertigo, oculogyric crises.</td>
</tr>
<tr>
<td>Barbiturates/benzodiazepines</td>
<td>Gaze-evoked nystagmus, saccadic pursuit, impairment of optokinetic nystagmus, slow saccades, vertical gaze palsy, external ophthalmoplegia, impairment of vestibulo-ocular reflex (VOR), internuclear ophthalmoplegia, central positional nystagmus, impaired vergence, decreased accommodative convergence, accommodation ratio.</td>
</tr>
<tr>
<td>Alcohol</td>
<td>Latent or persistent positional vertigo, saccadic pursuit, slow saccades, downbeat nystagmus, vertigo.</td>
</tr>
<tr>
<td>Benzodiazepines</td>
<td>Saccadic pursuit, slow saccades, impairment of VOR, impairment of gaze-holding.</td>
</tr>
<tr>
<td>Tricyclic antidepressants and L-tryptophan</td>
<td>Internuclear ophthalmoplegia, external ophthalmoplegia, opsonicnus.</td>
</tr>
<tr>
<td>Bromides</td>
<td>Ocular oscillations and myoclonus, internuclear ophthalmoplegia.</td>
</tr>
<tr>
<td>Vestibular sedatives</td>
<td>Impairment of VOR, slow saccades, alternating skew deviation, opsonicnus, downbeat nystagmus, vertigo.</td>
</tr>
<tr>
<td>Lithium</td>
<td>Opiocnus.</td>
</tr>
<tr>
<td>Thalidomide</td>
<td>Internuclear ophthalmoplegia, oculogyric crises.</td>
</tr>
<tr>
<td>Mehtaldrone</td>
<td>Saccadic pursuit, saccadic hypotrichia.</td>
</tr>
<tr>
<td>Haloperidol</td>
<td>Opiocnus.</td>
</tr>
<tr>
<td>Marijuana</td>
<td>Gaze-evoked nystagmus, saccadic pursuit, increased accommodation convergence, accommodation ratio.</td>
</tr>
<tr>
<td>Amphetamine</td>
<td>Spontaneous and/or positional nystagmus, impairment of optokinetic nystagmus.</td>
</tr>
<tr>
<td>Mercury</td>
<td>Vertebrobasilar hearing loss.</td>
</tr>
<tr>
<td>Chemotherapeutic antineoplastic agents</td>
<td>Transient vertebrobasilar hearing loss, impaired VOR.</td>
</tr>
<tr>
<td>Cyclophosphamide (ethylenimine, husemide)</td>
<td>Temporary vertebrobasilar hearing loss.</td>
</tr>
<tr>
<td>Aminoglycoside antibiotics (gentamicin, streptomycin)</td>
<td>Transient vertebrobasilar ocular impairment.</td>
</tr>
<tr>
<td>Acetylsalicylic acid</td>
<td>Central positional nystagmus, exaggerated VOR, impaired fixation suppression of VOR.</td>
</tr>
</tbody>
</table>

cupulolithiasis ("heavy cupula") is valid, then it remains unclear why patients with BPPV do not suffer from additional positional vertigo/nystagmus. Similarly, compounds differing in specific weights such as alcohol or heavy water should be able to induce not only positional but also positioning nystagmus with rapid changes in head position. In the case of alcohol this positioning nystagmus (positional alcohol vertigo/nystagmus (PAN)) should beat toward the same direction as positional nystagmus during the resorption phase (PAN I, when the cupula is relatively lighter) and toward the opposite direction during the reduction phase (PAN II, when the cupula is relatively heavier). Further experiments are needed in order to clarify these discrepancies between theory and clinical manifestation.

**Positional Alcohol Vertigo/Nystagmus**

Barany described the direction-changing characteristics of PAN in humans with changes in head position (beating toward the unanesthetized ear), which was later proved in animals. Walde described that the direction of PAN reverses (beating toward the uppermost ear) hours after alcohol intake. This phenomenon was later termed PAN II by Aschan et al. and Money et al. Peripheral labyrinthine origin of PAN was suggested by observations that it does not occur after loss of labyrinthine function in humans and in animals. This argument, however, is not convincing because central positional nystagmus is also dependent on peripheral graviceptive input.

**FIG.7–9.**

Ingestion of non-soluble molecules with differing specific weights such as alcohol, heavy water, or glycerol causes the specific gravity differential between cupula and endolymph (buoyancy hypothesis) with positional nystagmus and vertigo. During the resorption phase of alcohol, nystagmus beats toward the uppermost ear (PAN I), with the cupula relatively lighter than endolymph. Positional nystagmus beats toward the uppermost ear during alcohol-reduction phase (PAN II) as well as in glycerol, heavy water, and macroglobulinemia-induced positional nystagmus with the cupula relatively heavier than endolymph. The gravity-dependent deflection force on the cupula (D in insert) must be greater than the physiologic restoring force (C in insert) in order for the positional nystagmus to last as long as the precipitating head position is maintained.
Alcohol is lighter than endolymph, and when blood levels approach 40 mg/dL, alcohol diffuses into the cupula, rendering it lighter than endolymph, thereby transforming the semicircular canals into gravity-sensitive receptors.\textsuperscript{15} Nystagmus and vertigo then occur when the subject lies down. In phase I of PAN, the nystagmus bears toward the undermost ear (Fig 7–9; Table 7–7). With time, blood alcohol diffuses into the endolymph, equalizing its specific gravity to that of the cupula. There is then a "silent (intermediate) period\textsuperscript{21}, beginning between 3.5 and 5 hours after cessation of alcohol ingestion, when positional vertigo is absent. Alcohol selectively diffuses out of the cupula before it leaves the endolymph. This causes the cupula to be transiently denser than the endolymph, thus initiating phase II of PAN, which begins between 5 and 10 hours after cessation of drinking when there is a falling blood level at about 20 mg/dL. In PAN II, nystagmus bears to the uppermost ear.

Positional vertigo may persist until all alcohol eventually leaves the endolymph (equalizing the specific gravities of the endolymph and the cupula), but this may not transpire until many hours after the blood alcohol level has reached zero. PAN II is usually associated with motion sickness and is a major concomitant of the hangover. The "morning after" drink of alcohol may indeed reequalize the specific gravities and lessen, at least transiently, the upright symptoms.\textsuperscript{24}

### Positional "Heavy Water" Nystagmus

Money and Myles\textsuperscript{25} described ingestion of 100 to 200 g deuterium oxide, which caused a vigorous lateral positional nystagmus lasting some hours in man, with a directional characteristic opposite to that of postural alcohol nystagmus. Deuterium oxide ("heavy water") has a molecular weight of 20.030 (that of water is 18.016), and is thought to diffuse farther into the cupula than the endolymph. As long as a great enough specific gravity deferential is maintained between the two, the cupula may act inappropriately as a gravity transducer.

### Positional Glycerol Nystagmus

The standard doses of glycerol used for obtaining diagnostic audiograms in patients with suspected Meniere's disease can cause transient positional nystagmus. This was first observed in a sin-

| TABLE 7–7. Positional Alcohol Nystagmus (PAN) |
|-----------------|-------------------------------|-----------------|
| **Phase**       | **nystagmus**                 | **Time**        | **Mechanism** |
| PAN I (reappraisal phase) | Direction changing rotational vertigo and nystagmus with head right or left, pointing toward the undermost ear | 30 min after oral administration; duration, 3 to 4 hr | With blood levels of 25 to 40 mg/100 mL, diffusion into the cupula makes alcohol lighter than endolymph, and sensitive to gravity changes |
| "Silent (intermediate period)" | No positional vertigo/nystagmus | 3 to 5 hr after alcohol ingestion | Alcohol diffuses also into the endolymph equalizing specific gravity of cupula and endolymph |
| PAN II (reduction phase) | "Hangover vertigo" with direction changing positional nystagmus with head right or left, toward the uppermost ear | 5 to 10 hr after alcohol ingestion | Alcohol stays longer in the endolymph, causing a specific gravity differential with the cupula being "heavier" |
gle case by Angelborg et al.\textsuperscript{7} and later studied more systematically by Rietz et al.\textsuperscript{9,10} In their study, five of six subjects exhibited a positional nystagmus that was maximal 1:30 minutes after glycerol ingestion, shortly after peak serum levels were achieved. From their data, Rietz et al.\textsuperscript{10} inferred that different transport velocities of the compound result in a postural nystagmus toward the uppermost ear. This concept is supported by animal studies in guinea pigs in which Yoshida et al.\textsuperscript{11} found that endolymphatic pressure dropped 15 minutes after intravenous administration of glycerol (when glycerol enters the cupula?) lasting for at least another 25 minutes, at which point glycerol entering the endolymph rebalances the osmotic gradient. The duration of postural glycerol nystagmus, most pronounced between 90 and 240 minutes after oral ingestion,\textsuperscript{12} indicates that glycerol does not enter the endolymph for at least 2 hours after oral administration.

**Positional Nystagmus With Macroglobulinemia (Waldenström’s Disease)**

Malignant lymphoproliferative Waldenström’s disease with macroglobulinemia can have auditory and vestibular manifestations in as many as 10% to 20% of those afflicted.\textsuperscript{42, 89} Various explanations of these manifestations have been proposed: increased blood viscosity with obstruction in the venules,\textsuperscript{8} stagnation or sudden release of clotting factors,\textsuperscript{54, 107} associated neurologic dysfunction,\textsuperscript{97} or hemorrhage.\textsuperscript{4} Vascular vertigo in the hyperviscosity syndrome is most likely due to obstruction of the venules and capillaries with peripheral vestibular hypofunction.\textsuperscript{3} This mechanism does not explain positional vertigo in Waldenström’s disease.

Keim and Sachs\textsuperscript{23} reported on 5 patients, 3 of whom gave a history of periodic dizziness with postural changes. They were able to record a direction-changing positional nystagmus with a latent period and fatique in 1 patient, who simultaneously reported rotary vertigo with nausea and visual disturbance. The 2 others, symptom-free at the time of otorhinological investigation, did not exhibit positional nystagmus. Because molecular weights in macroglobulinemia of over one million are possible, as opposed to normal gamma globulin values with a molecular weight of approximately 150,000, Keim and Sachs stress the increased specific gravity of the cupula as the causative factor, which varies with concentration of the circulating protein and the diffusion characteristics.

**Central Positional Vertigo**

When the position of the head is brought to an off-vertical, lateral, or head-hanging position a change in graviceptive (otoletic) input occurs. This change is the precipitating factor for central positional vertigo. The most probable explanation for this response is a vestibular tone imbalance with directional positional nystagmus and rotational/linear vertigo caused by disinhibition of the vestibular responses on perception, eye, head, and body position.\textsuperscript{27} Thus, it is not (as one might speculate) the head position—dependent dislocation of the mass and intracranial structures which causes the manifestations.

There are a variety of central positional vertigo syndromes resulting as a consequence of a mass (tumor, hematoma) near the fourth ventricle and the vestibular nuclei (Fig 7–10). These are characterized by rotational vertigo, nystagmus, and postural imbalance, which may be abrupt and more violent than in peripheral labyrinthine dysfunctions. This severe form of central positional vertigo, which initially immobilizes the patient, gradually improves within days to weeks. The central vestibular pathways involved are not yet known. It is known, however, that the typical site of the lesion is dorsolateral to the fourth ventricle.
Positional nystagmus without concomitant vertigo is usually central; however, the direction of the nystagmus varies. It may beat diagonally or toward the undermost or uppermost ear. Frequently it is bilateral and direction changing when the head is tilted toward the right or left, respectively.

The frequency of central positional nystagmus is usually low and constant, which distinguishes it from BPPV. Differentiation between central positional nystagmus and BPPV is based not only on the direction of the nystagmus but also on the lack of a latency period after movement to the provoking position. In addition, there is a lack of fatigability and habituation on repetitive stimulation.

Central positional nystagmus is indicative of a posterior fossa lesion. The probable locations are the caudal brain stem and the vestibulocerebellum; however, the condition does not allow for a more precise localization. Computed tomography and magnetic resonance imaging are also insufficient in determining the location of the lesion. The possible causes are similar to those of positional downbeating nystagmus, and we have also seen it with rapidly developing hydrocephalus if it involves the fourth ventricle. It is our experience that central positional nystagmus occurs frequently in the elderly (lacunar ischemia?) and often recovers spontaneously (central compensation?).

There is some evidence, from experiments in animals, that posterior cerebellar vermis lesions may cause positional nystagmus which mimics BPPV. The clinical relevance of this central "pseudo-BPPV" has been repeatedly stressed[10, 12]; however, it should not be overestimated because of its rarity. Other reports that Borrelia infections are a possible causative factor for both peripheral and central positional nystagmus[13] still need to be proved and analyzed more thoroughly.

Positional Downbeating Nystagmus

Positional downbeating nystagmus, with only slight vertigo with the patient in the head-hanging position, is indicative of a vestibulocerebellar nodular lesion. It may be related to the downbeat nystagmus syndrome, which also shows activation on head extension. Experimental extirpation of the nodulus in the cat causes postural downbeat nystagmus.[12] This has also been confirmed by clinical experience[13, 14]. Physiologically,
the nucleus may have an inhibitory influence on the gain of the VOR. Thus, vestibular vertigo and ataxia and drop attacks can be undoubtedly precipitated by extreme extension or rotation of the neck, at least when a partial obstruction of the arterioles combines with a sudden fall of systemic blood pressure, for example, in a patient rising from a chair and looking up. An association of dysfunctions that convinces the clinician in the diagnosis is that of vertigo with visual illusions, field defects, diplopia, dysphagia, dysarthria, drop attacks, or motor symptoms.

**Basilar Insufficiency**

Nystagmus, vertigo, and postural imbalance can be caused by multiple sclerosis, ischemia, intoxication, cranio-cervical malformation, or cerebellar degeneration; however, sometimes there is no identifiable cause in elderly patients. Brain imaging techniques in most cases do not show a vestibulocerebellar lesion.

Apart from basilar insufficiency, however, symptoms of tachycardia and postural imbalance occur frequently in healthy people such as that elicited by overhead work while standing on an unstable wobbling ladder or in situations in which visual cues conflict with proprioceptive input (looking up at moving clouds).

Vertigo and postural imbalance terminate abruptly when the head is flexed to a neutral position. The symptoms are also often attributed to intermittent vertebral artery occlusions caused by the head posture, particularly in the elderly. The symptoms, however, occur frequently in young people, and a physiologic explanation is presented based on an unusual combination of multisensory inputs from the stabilizing systems. The "normal" instability related to this head position can be determined by attempting to balance on one foot with the eyes closed and head extended as compared with the neutral head position. The otoliths are beyond their optimal functioning range in the offending head-extended position. When the body is supine, the otoliths are in the same position relative to the plane of gravity but are not, in this situation, involved in postural control. The otoliths are also out of optimal range with head flexion, but vertigo is not induced. Flexion, however, is a common position.
**OTHER POSITIONAL/POSSESSION TITLES**

Head Motion Intolerance With Bilateral Vestibulopathy

The characteristics and origins of bilateral vestibulopathy are summarized in Table 7-8. Bilateral vestibular loss causes unsteadiness of gait and oscillopsia associated with head movements or when walking. This is due to an inappropriate VOR, and excessive motion of images on the retina was measured in these patients while walking.57 The condition can be identified by a simple bedside test of the VOR (see Fig 7-2) as well as by the decreased ocular motor responses to thermal irrigation and angular acceleration.

Some ototoxic drugs (for example, streptomycin, gentamicin) are known to damage the peripheral vestibular sensory cells in advance of those in cochlea (hair cell damage of the inner ear). Kanamycin, tobramycin, and neomycin preferentially damage the auditory sensory cells,18 whereas more recent ami-

### TABLE 7-8.

<table>
<thead>
<tr>
<th>Syndrome</th>
<th>Causes</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Clinical syndrome</strong></td>
<td><strong>Symptoms</strong></td>
</tr>
<tr>
<td>Unsteadiness of gait (particularly in dark)</td>
<td>Oculocollis with head movements (e.g., when walking)</td>
</tr>
<tr>
<td><strong>Signs</strong></td>
<td></td>
</tr>
<tr>
<td>Gyroparetic vestibulo-ocular reflex bedside test</td>
<td></td>
</tr>
<tr>
<td><strong>Etiology</strong></td>
<td></td>
</tr>
<tr>
<td>Otic pathy</td>
<td></td>
</tr>
<tr>
<td>Labyrinthine infections</td>
<td></td>
</tr>
<tr>
<td><strong>Bilateral sequential vestibular neuron</strong></td>
<td></td>
</tr>
<tr>
<td><strong>Tinnitus (neuromastomatosis)</strong></td>
<td></td>
</tr>
<tr>
<td><strong>Endolymphatic hydrops</strong> (delayed endolymphatic hydrops)</td>
<td></td>
</tr>
<tr>
<td><strong>Hydrops</strong></td>
<td></td>
</tr>
<tr>
<td><strong>Autoimmune inner ear disease</strong></td>
<td></td>
</tr>
<tr>
<td><strong>Gliosis</strong></td>
<td></td>
</tr>
<tr>
<td><strong>Tight's disease</strong></td>
<td></td>
</tr>
<tr>
<td><strong>Congential malformation</strong></td>
<td></td>
</tr>
<tr>
<td><strong>Familial vestibular areflexia</strong></td>
<td></td>
</tr>
<tr>
<td>**Vertebrobasilar ischemia ( ?)</td>
<td></td>
</tr>
<tr>
<td>**Vestibular ataxias ( ?)</td>
<td></td>
</tr>
<tr>
<td>**Vestibular aging ( ?)</td>
<td></td>
</tr>
<tr>
<td>**Idiopathic bilateral vestibulopathy ( ?)</td>
<td></td>
</tr>
</tbody>
</table>

**Management**

Controlled physical exercise consisting of voluntary head movements about different axes when fixing a target, and balance training (recruitment of compensatory nonvestibular control capacities, somatosensory and visual)


**Note:**

- Some ototoxic drugs like dihedral and ribosorbic acid are also used.
- Hair cells and cochlear neurons may be transiently (reversibly) damaged by diuretics (ethacrynic acid, furosemide) or high-dose salicylate therapy.18,113 Combined use of loop-inhibiting diuretics and aminoglycoside antibiotics can cause permanent hearing loss. Permanent loss of hair cells sometimes occurs with alkylation antineur chemotherapeutics.
- Independent of these produced by ototoxicity, single cases of "progressive vestibular degeneration" of unknown cause have been described with the following factors in ear: repeated episodes of dizziness relatively early in life, bilateral loss of vestibular function with...
retention of hearing, and freedom from other neurologic disturbances.16,22,38

Bilateral vestibulopathy may occur for other more common reasons such as meningitis, labyrinthine infection, bilateral tumors (Fig 7-11), end-lymphatic hydrops, inner ear autoimmune disease, or congenital malformation.32,119 Autoimmune disease of the inner ear can produce both vestibular and auditory loss.99 Permanent bilateral loss of vestibular function has also been described for bilateral sequential vestibular neurectomy.173 There are, however, a considerable proportion of patients presenting with seemingly idiopathic bilateral vestibulopathy.12 There is reason to assume that these patients reflect a heterogeneous group of causes, some of which may be vascular.

Management

Controlled physical exercises, including voluntary head movements about all three axes and balance training, can improve the condition in patients with permanent bilateral vestibulopathy. This will recruit nonvestibular sensory capacities such as the cervico-ocular reflex or proprioceptive (somatosensory) control of stance and gait. The latter process of multisensory compensation is indirectly demonstrated by the significantly higher gain of cervico-ocular reflexes and autokinetic nystagmus in labyrinthine-defective subjects.18

Clinical Evidence for Cervical Vertigo?

The frequency and intensity of cervical vertigo should not be overstated. Pain arising from the cervical spine with tenderness and limitation of movement of the neck, if associated with iatrogenic vertigo and nystagmus, should not be called cervical vertigo. The characteristic symptom of cervical vertigo (Table 7-9) is more likely to be a sema-

FIG 7-11.
Magnetic resonance imaging of a patient suffering from neurosphenepritis. Axial (A) and frontal (B) orientation after application of intravenous gadolinium-DTPA as a contrast agent with echo delay time (TE), 17 ms; pulse sequence repetition time (TR), 552 ms. Multiple neuritis involve cranial nerves bilaterally. A, there is an infratemporal acoustic neuritis (arrow) as well as hypothalamic neuritis. B, 3 years after surgical removal of an acoustic neuritis, an extratemporal neuritis has recurred (arrows) pressing on and obstructing the pontine brain stem. (Courtesy of Dr. Voigt, Department of Radiology, University of Munich.)
tion of numbness or floating with ataxia of stance and gait, as can be inferred from the experimental vertigo induced by unilateral suboccipital local anesthesia.35 It is not known how traumatic, degenerative, inflammatory, or rheumatic disease affect neck sensory input. This explains the various hypotheses, for example regarding cervical vertigo following whiplash injury. Suggested causes are the neuromuscular mechanism,44 or mechanical vascular obstruction of the vertebral artery.33 The incidental observation of an improvement of post-traumatic vertigo and ataxia by use of a neck collar was made earlier by Longen48 in 1845. But head trauma and whiplash injury do not affect only neck structures. The otoliths are obviously more vulnerable to acceleration; damage causes otolith vertigo with a similar benign course to that of neck pain.42 Furthermore, whiplash injuries frequently damage the brain,107 which makes interpretation of an abnormal vestibulo-ocular test difficult.40,127 Postural testing with head extension as proposed by Defong and Bles,44 or nystagmus induced by passive head rotations90,98,111 cannot clearly differentiate between healthy and diseased subjects.

Vertigo and/or Tinnitus Associated With Neurovascular Compression ("Disabling Positional Vertigo," "Vestibular Paroxysmia")

Neurovascular cross-compression of the root entry zone of the cranial nerves V, VII, and IX can cause local demyelination and axonal hyperactivity (for example, by transversely spreading ephaptic activation), accompanied by the distressing symptoms of trigeminal neuralgia, hemifacial spasm, and glossohypoglossal neuralgia. Compressing vessels which had made indentations on the relevant nerve or nerve root entry zone have been found surgically, and the efficacy of microvascular decompression as a successful treatment of these symptoms is well established.24 These findings suggest that it is reasonable to search for a group of patients presenting with typical paroxysmal vestibular and/or cochlear symptoms, analogously caused by neurovascular compression of the eighth cranial nerve.

If vertigo is the major complaint, vascular compression must be causing abnormal impulse activity of the vestibular portion of the eighth cranial nerve. If vertigo and tinnitus are present, then both vestibular and cochlear portions of the nerve are compressed by one or more vessels, either the anterior inferior cerebellar artery (AICA), the posterior inferior cerebellar artery (PICA), or a vein. Compression may be due to vascular malformation of the posterior fossa,30 arterial ectasia,132 or simply to arterial aging with elongation and looping. It is well established that pulsatile compression of the caudal cranial nerves is more likely to be symptomatic when the central (oligo-)dendroglia) myelin rather than the peripheral myelin is involved.
For the eighth cranial nerve this means that the entire intracranial portion from brain stem to the internal auditory canal (1 to 1.5 cm) may be particularly vulnerable. This syndrome was first described by Jannetta et al.73 and later termed 'disabling positional vertigo' by the same authors.74,75 A description for a most heterogeneous collection of signs and symptoms, and far from a reliable diagnosable disease entity. The lack of a well-defined syndrome, and of a diagnostic test, make it difficult for the nonsurgical clinician to "believe in" this interesting disease. However, it is possible that it plays an important role in the group of vertigo patients who do not respond to other treatment.

Hesitation in the diagnosis of this disorder is justifiable because retro-mastoid craniotomy and microvascular decompression of the eighth cranial nerve is the recommended procedure once the diagnosis has been established. This surgical procedure is still associated with a mortality of about 1% and a considerable morbidity of about 10%.76,77 Even though others report lower mortality/morbidity rates, selection criteria for the operation that have been proposed are insufﬁcient, because they do not reliably exclude causes of vertigo other than neurovascular compression. Too many patients will be incorrectly selected for an unnecessary and risky operation. The surprisingly high improvement rate (16 of 21 patients became symptom free after operation)78 is not signiﬁcant proof of the diagnosis, since other common vertigo syndromes such as "phobic postural vertigo" can be treated rapidly and successfully by psychiatric methods. Furthermore, it is necessary to remember that decades passed before it was demonstrated that the success of "endolymphatic sac surgery" operations in the management of Meniere's disease was a result of a placebo effect.79 This is the reason why the following description of the so-called "disabling positional vertigo" should be accepted with critical reserve.

In their original paper, Jannetta et al.73 reported on 9 patients, all of whom showed vascular compression of the superior or inferior vestibular nerve (or both) at the brain stem, and 8 of whom experienced total relief of symptoms after microvascular compression.

"We have found a certain kind of balance disorder that does not fulﬁll the criteria of the established syndromes just discussed (Meniere's disease, benign paroxysmal positional vertigo, vestibular neuritis) and that does not respond to medical treatment. Patients with this disorder experience a constant positional vertigo or disequilibrium so severe that they are disabled and constantly nauseated; thus, the vertigo cannot be called benign."75

A more recent description of the syndrome in another 21 patients is even less speciﬁc and more ambiguous. To complicate matters and further stretch the clinical spectrum of neurovascular compression syndromes, Moller72 reports on 10 patients, all of whom had classical symptoms of Meniere's disease, but who, in addition, had audiologic signs indicating involvement of the auditory nerve. Because vertigo is so common does not allow identiﬁcation of the site of the lesion, results of audiovestibular tests and brain stem auditory evoked potentials (BAEP) served as the basis for deciding which was most probably the affected side and the side that would eventually be subjected to surgical exploration. Those BAEP recordings with unilaterally increased latencies between the waves I and III were reported to be the most helpful. These patterns are similar to those seen in acoustic neuromas and indicate functionally signiﬁcant lesions of the proximal cochlear portion (waves I and II) of the eighth cranial nerve.

First reports from other clinical centers do not clarify the middile of signs and symptoms seemingly characteristic for this mysterious disease. Of 10 pa-
tients diagnosed as having neurovascular compression of the eighth cranial nerve, all were middle-aged and hypertensive, and most complained of paroxysmal vertigo lasting from hours to days, which was aggravated by changing head position (movement). Also, patients had persistent tinnitus and progressive unilateral hearing loss. In contrast to the findings of Jannetta et al.25 and Muller et al.49 there was no positional vertigo, and wave I of BAEP was prolonged in 6 cases, whereas the interpeak latency of waves I to III was normal in 5 cases.

In conclusion there is some evidence that neurovascular compression of the eighth cranial nerve can cause paroxysmal attacks and a persistent disabling vertigo/binaural syndrome (mostly with tinnitus and some hearing loss) and that this can be successfully treated by microvascular decompression procedures.

The proposed name of the disease, “disabling positional vertigo,” is misleading. “Vertibular paroxysmia” might be a more appropriate term.25 It is not only the position of the head but the movement (change in position) that seems to aggravate vertigo and postural imbalance in many patients. Neither the regular surgical finding of close vessel/nerve contact at the brain stem, nor the surprisingly high improvement rate following microvascular decompression unequivocally confirm the proposed diagnosis.

There is no pathognomonic sign for vestibular paroxysmia, and many elderly patients will exhibit some unilateral or bilateral deficit of cranial nerve function which may easily persuade the unscrupulous clinician to undertake an unnecessary posterior fossa craniotomy. Antivertigo drugs have been reported to be ineffective in these patients, but there is no evidence that antiepileptic drugs, such as carbamazepine, have been thoroughly tested. Carbamazepine, the drug of first choice for trigeminal neuralgia, was indeed effective in a few vertigo patients in whom we suspected a neurovascular compression syndrome of the eighth cranial nerve. Further information is required if the clinical picture presented by the eighth cranial nerve neurovascular compression syndrome is to be accurately defined.

CONCLUSIONS

Benoia paroxysmal positioning vertigo due to cupulolithiasis or canalolithiasis is the most important disorder described in this chapter because it is the most common cause of vertigo in the elderly. Brief attacks of rotational vertigo and concomitant positioning-induced rotatory-linear nystagmus are precipitated by rapid extension of the head or by lateral head tilt toward the affected ear. Definitive diagnostic nystagmus criteria are latency of 1 to several seconds, beating direction rotating toward the unaffected ear, duration of less than 1 minute, reversal of nystagmus direction with return of the patient to the upright position, and fatigability with repetition of the maneuver. Spontaneous recovery is common, but there is a highly effective form of mechanical therapy that involves the use of positioning maneuvers.

Positional alcohol (vertigo) nystagmus, when elicited by lateral head position, beats toward the undermost ear during the resorption phase of alcohol (PAN I) and toward the uppermost ear during the reduction phase (PAN II). It can be explained by the buoyancy hypothesis: A specific gravity differential between cupula and endolymph occurs when alcohol (which is lighter than endolymph) diffuses into the cupula, thereby transforming the semicircular canals into gravity-sensitive receptors.

Until recently there are no reliable clinical tests available by which to establish the diagnosis of cervical vertigo following whiplash injury, or neurovascular cross-compression (“vestibular paroxysmia”). Most cases of transient post-traumatic vertigo that manifest with head motion intolerance and un-
Usefulness of Positional and Positioning Testing

steadiness of gait may be otolith vertigo. Central compensation (rearrangement) would account for the gradual recovery within days to weeks, supporting the view that exercise is the best therapy.

General positional vertigo and/or nystagmus are disorders of intracranial posterior fossa structures, particularly when involving the vestibular nuclei and/or connecting pathways to the vestibulo-cerebellum. These disorders are often associated with other otoclonal abnormalities such as sacculo-striate pursuit, gaze-evoked nystagmus, and upbeat or downbeat nystagmus. Positional nystagmus without vertigo is always central in origin. But lesions dorsolateral of the vestibular nuclei may cause a severe form of position-dependent vertigo that includes nausea and vomiting. A lesion of toxic or due to the inhibition of (inhibitory) vestibulocerebellar Purkinje cell input onto vestibulocerebellar nuclei may be causative.

Positional and positioning tests may be contaminated by spontaneous vestibular nystagmus secondary to a perilyncal (vestibular neuritis, Meniere's attack) or central (AICA or PCA infarction; Wallenberg's syndrome) vestibular tone imbalance. Spontaneous vestibular nystagmus is typically suppressed by fixation. Voluntary oscillatory saccades which override fixation, such as congenital nystagmus or downbeat or upbeat nystagmus, may mimic positional nystagmus in that they are modified in intensity by gaze direction (congenital nystagmus) or by static head tilt to the prone or supine position (downbeat nystagmus).

REFERENCES

runs in the cat. Exp Brain Res 1986; 63:
35–48.


64. Harris CS, Gouldby JE, Graybiel A: Positional Alcoholic Nystagmus in Relation to Labyrinthine Function. NSAM 839, NASA 1947. Ponsarnau, Fig. Naval School of Aviation Medicine, 1962.

65. Harrison MS, Onzahhigah C: Positional ver-

66. Hasegawa T: Die Veränderung der labri-

67. Herdman SJ: Treatment of benign parox-


70. Hulse M: Die zentrale Gleichgewichtsstörun-

71. Hydek G: Extra-cranial circulatory compli-

72. Iyeruji M, Nagaya M: Vestibular endolymphatic drainage in squirrel monkeys after exposure to intense linear acceleration. In: Third Symposium on the Role of the Vestibular Or-

73. Jarrett PF: Neurocerebral cross-

74. Janetta PJ: Treatment of trigeminal neural-

75. Janetta PJ, Meller MD, Moller AR: Disabil-

76. Johnson LG, Hulse J: Sensory and neural 
degeneration with aging, as seen in mi-

77. Jung R: Nystagmusregele, Zur Physiologie und Pathologie des ortsspechendem vestibulum Ver-
tens beim Mensch, in von Bergmann G, Frey W, Schwieb W (eds): Handbuch der In-

78. Karlici T: Der biphausische undendurende Kopf-

79. Kasatkana A: Nystagmus of positional para-

80. Kasatkana A, Kerkkam T: Parovisual posi-
itional vertigo: A study of 235 cases. J Oto-

81. Kellah CJ, Kolody MP, Lienswobd AJ: Posi-


83. Lindsay JR, Hemnesweg WG: Postural ver-
85. Lacelle-Dufour J, Silverstein P, Coe J: Neurological aspects of Waldenström's macroglobu-

86. Longe JA: Mikrotomie sur les troubles qui surviennent dans l'équilibre, la station et l'actionnement des animaux après la sec-

87. Longridge NS: Barlow HD: Bilateral parasy-

88. Mahrman CH, Diamond SG, Juicke I: Intrac-
ural dysfunction in benign positional pos-
tional vertigo. In Graham MD, Keenan L (eds): The Vestibular System: Neuropa-

89. McClure JA: Horizontal canal BPPV. J Oto-
laryngol 1985; 14: 30-36.


92. Moller MD: Controversy in Meniere's dis-
 ease: Results of microvascular decom-

93. Moller MD, Alferfer AJ, Jannett PJ, et al: Diagnosis and surgical treatment of dis-


95. Money KE, Miles WS: Heavy water nystag-


97. Moore TP: Cerebellar microvascular decom-


99. Myers E, Bernstein J, Fasitopulos G: Sulli-


102. Patterson JS, McClure JA: Posterior semicir-
cular occlusion for intractable benign positional pos-


104. Romo ML, Roje CR, Ronis BJ: Otologic manifestiations of Waldenström's macroglobu-


106. Röthfeld J: Uber den Einfluss akuter und chronischer Alkoholvergiftung auf die Funk-

107. Ruben JR, Drenfield A, Berg P: Sudden se-
quelae of aneurysm as the presenting symptom of macroglobulinemia. JAMA 1960; 209: 1364-1365.

108. Rubini W: Whiplash with vertebrobasilar involve-


110. Saio K: Histopathological study on the vestib-
ular toxicity of six amnimumephrine antibiot-


112. Schmidt CL: Zur Pathophysiologie des pe-


114. Schluens HF: Capululthikisch. Arch Oto-

115. Schluens HF: Pathology of the Ear. Cam-


117. Schluens HF, Witt JR: Acute bilateral se-
quelatory vertigo in cirrhosis. Am J Oto-


