

BEDSIDE NEURO-OTOLOGICAL EXAMINATION AND INTERPRETATION OF COMMONLY USED INVESTIGATIONS

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J Neurol Neurosurg Psychiatry 2004;75(Suppl IV):iv32–iv44. doi: 10.1136/jnnp.2004.054478

The assessment of the patient with a neuro-otological problem is not a complex task if approached in a logical manner. It is best addressed by taking a comprehensive history, by a physical examination that is directed towards detecting abnormalities of eye movements and abnormalities of gait, and also towards identifying any associated otological or neurological problems. This examination needs to be mindful of the factors that can compromise the value of the signs elicited, and the range of investigative techniques available. The majority of patients that present with neuro-otological symptoms do not have a space occupying lesion and the over reliance on imaging techniques is likely to miss more common conditions, such as benign paroxysmal positional vertigo (BPPV), or the failure to compensate following an acute unilateral labyrinthine event.

The role of the neuro-otologist is to identify the site of the lesion, gather information that may lead to an aetiological diagnosis, and from there, to formulate a management plan.

BACKGROUND

Balance is maintained through the integration at the brainstem level of information from the vestibular end organs, and the visual and proprioceptive sensory modalities. This processing takes place in the vestibular nuclei, with modulating influences from higher centres including the cerebellum, the extrapyramidal system, the cerebral cortex, and the contiguous reticular formation (fig 1). Therefore any derangement of the structure or function of the sensory inputs, the central vestibular structures or the effector pathways—that is, the oculomotor and vestibulo-spinal pathways—is likely to result in a balance disorder.

Many conditions will elude diagnosis if balance is equated purely with a disorder of vestibular function. Drachman and Hart¹ have emphasised the importance of multi-sensory dizziness, particularly in the elderly, when two or more of the following conditions are present: visual impairment, peripheral neuropathy, vestibular deficit, cervical spondylosis, and orthopaedic disorders affecting the large joints. An appreciation that dysequilibrium may be consequent on multiple pathologies is essential if the appropriate investigation and interpretation of the data are to be achieved.

General medical conditions similarly may contribute to dizziness—that is, postural hypotension, vasovagal syncope, cardiac valvar disease, hyperventilation—and a full and comprehensive history is vital. A general medical examination, with particular attention to the eyes, the ears, the nervous system, the cardiovascular system, and the locomotor system, may be indicated and general medical investigations should also be considered.

SECTION 1 Aural examination

A careful inspection of the ear is needed, as any otological pathology is likely to point the clinician to a peripheral rather than a central neuro-otological lesion. It includes examination of the auricle, the external auditory meatus, and the middle ear—that is, as far as can be assessed by examination of the tympanic membrane (TM). The tympanum offers a window into the middle ear cleft and is affected by most of the changes that can take place in the middle ear. Otitis externa, acute otitis media or TM perforation contraindicate caloric testing. Wax also precludes caloric irrigations because it acts as a heat seal, and also when impacted gives rise to a spurious conductive hearing loss.

Inspection of the ear may be carried out with the use of an otoscope or with a head-worn light source, leaving the hands free. The speculum should be directed around the circumference of the outer ear canal looking for debris, foreign bodies, inflammation, and for defects of the posterior or anterior wall. The magnifying lens of the Siegel's (or pneumatic) speculum can be fitted into the speculum and the bulb squeezed to raise intrameatal pressure and then relaxed, sucking the membrane outwards. In the presence of a middle ear effusion, the membrane is immobile; however with a very flaccid TM, this may have been sucked back onto the middle ear mucosa, and

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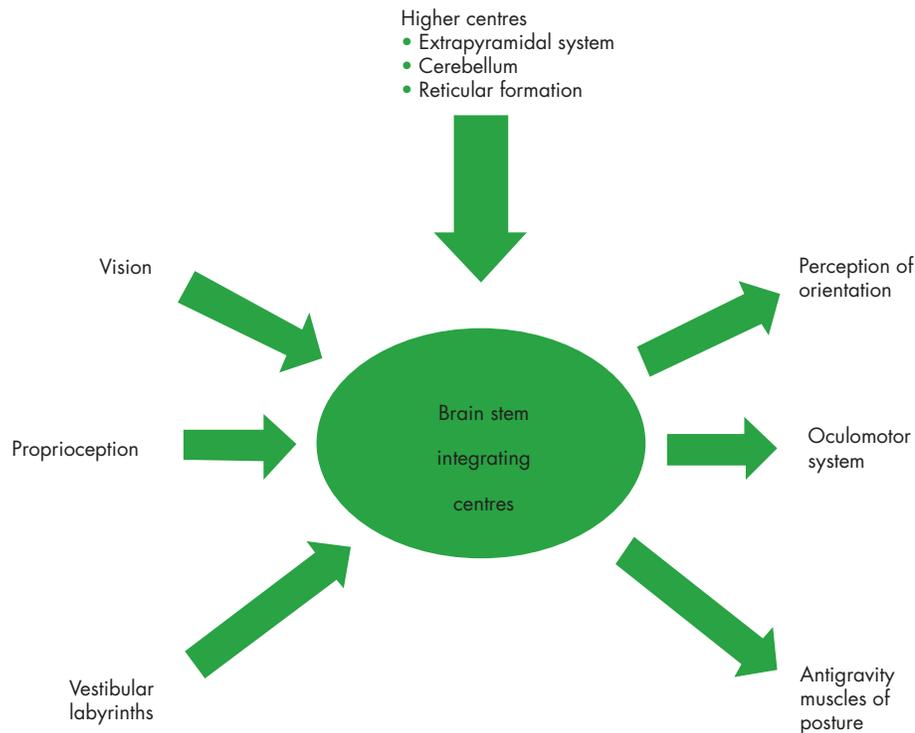


Figure 1 Sensori-motor physiology of the maintenance of balance showing the three sensory inputs required for maintenance of equilibrium, the central modulating influences, and the efferent pathways.

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lowering the pressure may suck it out again, allowing a retraction pocket to be distinguished from a perforation. Alternatively, the Valsalva manoeuvre—that is, auto-inflation of the ear—can be used to raise middle ear pressure and to observe similar changes.

Examination of the auricle (pinna)

The auricle is essentially vestigial, contributing only to collection of sound, slightly enhancing the efficacy of the ear. It should be inspected for signs of inflammation, trauma, surgical scars, or haematoma auris following a blow to the ear, and also for congenital deformities. Most developmental abnormalities are fairly obvious, but must be carefully identified because of the likely associated findings of middle and inner ear abnormalities—that is, anotia (absent auricle); microtia (smaller than normal and probably misshapen auricle). Pre-auricular appendages are found in 1.5% of the population; fistula auris, a small blind pit seen anterior to the tragus, results from incomplete fusion of the auricular tubercles.

Examination of the external auditory meatus

Congenital conditions include a stenosed or atretic external auditory meatus (EAM). In the latter, the EAM is closed over with a membranous, straight bony wall across the canal. This abnormality can be graded according to severity and radiological investigation needs to be pursued if found, to identify the size and shape of the middle ear cavity, and possibility of associated inner ear disorders.

Acquired abnormalities include:

- ▶ Foreign body obstruction
- ▶ Otitis externa. Likely infections: staphylococcus, pseudomonas and diphtheroids; fungal infections—that is, aspergillus and candida; and viral infections, identified by the vesicular eruptions of herpes zoster—that is, Ramsey-Hunt syndrome

- ▶ Osteomas are rounded excrescences of bone
- ▶ Exostoses are small osteomata and are quite common in people who swim or dive regularly.

Examination of the tympanic membrane and middle ear

Wax

If impacted, or obscuring the view of the tympanic membrane, the cerumen must be removed carefully without causing pain to the patient. Hard lumps can be removed

Syringing

Absolute contraindications to syringing

- ▶ The presence of an ear infection (otitis externa or media)
- ▶ The ear is known to have a perforation
- ▶ From the history, the ear is suspected to have a vulnerable tympanic membrane

Method

- (1) With the patient seated in a chair, protect him/her with a plastic cape and towel
- (2) Ask the patient to hold a kidney dish receiver on the shoulder, just below the pinna
- (3) Draw up in a metal syringe a solution of sterile water of 37°C (any variation from this will cause vertigo)
- (4) Draw the pinna upwards and backwards
- (5) Place the nozzle a few millimetres into the canal, pointing upwards and backwards
- (6) Direct the stream of water along the roof of the canal, between the skin and the wax

Note:

- ▶ Syringing should not be painful and should be stopped if the patient complains of pain
- ▶ With appropriate instruction, nursing staff can carry out this procedure
- ▶ Aural suction in the hands of an otologist may be required if the wax is not easily removed by the above measures

using the Jobson-Horn probe, or a Cawthorne wax hook. An alternative method for removing wax is syringing. If the wax seems very hard, it can be softened over a period of weeks using warm olive oil drops administered nightly, or alternatively 5% sodium bicarbonate drops, or a ceruminolytic preparation available commercially.

Tympanic membrane and middle ear

The standard landmarks of the tympanic membrane (TM) are: the central portion with handle of the malleus visible through the drum; the pars flaccida superiorly, and the pars tensa inferiorly; identification of the long process of the incus and the stapedius tendon seen in more TMs. The following are important features to identify:

- ▶ **Perforations**—when seen in the pars tensa, these are classified as marginal or central; defects of the attic portion are described as attic perforations; and the position of the perforation is defined as anterior, inferior, or posterior; large central perforations are described as subtotal and large marginal perforations as total.
- ▶ **Colour**—the normal eardrum has the appearance of mother of pearl. The light reflex is found antero-inferiorly, where reflection of the examination light occurs. If the membrane is thickened, the reflex may be lost. If the middle layer of fibrous tissue of the TM undergoes hyaline degeneration it may become impregnated with deposits of calcium—that is, tympanosclerosis.
- ▶ **Position of the membrane**—Retraction of the membrane occurs when there is chronic lowering of pressure in the middle ear—that is, chronic obstruction of the Eustachian tube. The handle of the malleus is drawn inward and there is retraction of the drum toward the medial wall of the middle ear. When severe, the drum is stretched around the long process of the incus and head of stapes, and at worst, the membrane is plastered against the promontory. (A fluid level may be visible if secretory otitis media has resulted from reduction of middle ear pressure. In the presence of air bubbles, middle ear fluid is confirmed.) With raised middle ear pressure, the drum may bulge outwards and depending on the colour, this increased middle ear pressure may be caused by acute suppurative otitis media (cherry red), or if a normal colour, the bulge is likely to be caused by raised air pressure in the middle ear alone.

Disorders of the tympanic membrane and middle ear

Congenital abnormalities include fusion of the ossicles—that is, congenital stapes fixation, absent stapedius tendon, and uncovered or aberrant VIIIth nerve. Acquired disorders include acute otitis media (fig 2), chronic otitis media, cholesteatoma, serous otitis media, and ossicular abnormalities.

Cholesteatoma is a cyst lined with squamous epithelium, which can arise in ears undergoing long periods of negative middle ear pressure and persisting middle ear infection—that is, chronic suppurative otitis media. Cholesteatomatous cysts are likely to begin in the attic of the ear and extend into the mastoid antrum. They are filled with cast-off epithelial cell debris and slowly increase in size. They can erode the surrounding bone and produce intracranial complications by eroding through the dura of the middle or posterior fossa, or through the lateral sinus or into the lateral semicircular canal (when a positive fistula sign would be elicited, see below). Cholesteatoma can be diagnosed from a history of perforation, chronic foul smelling discharge from the ear, and



Figure 2 Acute otitis media: otoscopic appearance of left tympanic membrane. Reproduced with permission of Professor Anthony Wright, Institute of Laryngology and Otology, London.

keratin debris in the pars flaccida area on otoscopic examination. It is potentially serious and requires surgical removal.

Tubo-tympanic disease describes chronic active otitis media unassociated with cholesteatoma. It is characterised by recurrent infections rather than persistent infections and by odourless discharge rather than offensive discharge. A central TM perforation and a break in the ossicular chain or malleus fixation are regarded as “safe” and unlikely to be associated with cholesteatoma.

Serous otitis media is recognised by an air/fluid level in the middle ear, or a bluish discolouration of the drum. A lack of compliance of the drum is found on tympanometry. Other effusions into the middle ear include blood (for example, haemo-tympanum after head trauma) or cerebrospinal fluid within the middle ear space.

Otosclerosis

This is an inherited, autosomal dominant hearing disorder, tending to present in later childhood/adulthood. Deposition of bone in the oval window niche occurs leading to fixation of the stapes footplate and a conductive hearing loss. The otosclerotic process can extend to involve the otic capsule, leading to an additional sensori-neural hearing loss and vertigo. Typically in the early osteoblastic phase, the appearance of the malleus head is hyperaemic—that is, Schwartze’s sign.

Glomus tumour

This is a jugulo-tympanic paraganglionoma which tends to expand within and traverse the petrous temporal bone by way of the pneumatized air cell tracts. It can present with pulsatile tinnitus, and a vascular mass lying behind the tympanic membrane can be identified (also described as the “setting sun” sign). These tumours may also extend into the labyrinth, or present as cranial nerve abnormalities.

TESTING OF AUDITORY FUNCTION

Tuning fork tests

Tuning fork tests have been traditionally used to distinguish conductive from sensorineural hearing loss and to identify functional hearing loss. With the advent of pure tone audiometry, only a few of these tests are still used clinically. The principles of tuning fork tests are:

Fistula sign

This is elicited in those patients where transmission of air pressure changes from the EAM is possible through a fistula into the labyrinth:

- ▶ raised pressure causes a conjugate deviation of the eyes towards the opposite ear and with maintenance of pressure, a corrective fast eye movement will be introduced.
 - ▶ the nystagmus will be towards the affected ear
- Depending on where the fistula has developed, the nystagmus will be:
- ▶ horizontal (horizontal semicircular canal)
 - ▶ torsional (anterior canal)
 - ▶ vertical (posterior canal)

EAM pressure may be raised by tragal pressure, but more accurately by tympanometry. Hennebert's sign is a positive fistula sign in the presence of an intact TM.

- ▶ the inner ear is more sensitive to sound conducted by air than bone
- ▶ in pure conductive hearing loss, the affected ear is subject to less environmental noise and is more sensitive to bone conducted sound.

The most commonly used tuning forks are those at 256 and 512 Hz. Lower frequencies produce a vibrotactile stimulus which can result in misleading thresholds. In practice the 256 Hz tuning fork better distinguishes air–bone gaps than the 512 Hz fork. When performed, the prong (tine) of the tuning fork should be struck against a hard, but elastic, mass (for example, a rubber pad) to prevent production of

Rinne's tuning fork test

Heinrich Rinne described his tuning fork test in 1855

- (1) The fork is struck and held with the tines perpendicular to the long axis of the external auditory meatus with the closest tine 1 cm from the entrance to the meatus
- (2) The patient is asked to report if he can hear the sound (AC)
- (3) The fork is immediately transferred behind the ear with the base firmly pressed to the bone overlying the mastoid (BC)
- (4) The patient is asked which sound is louder: that "in front of the ear", or that "behind the ear"

Positive Rinne test: if $AC > BC$ —that is, the sound in front of the ear is reported as louder:

- ▶ indicates normal hearing
- ▶ or an ear with a sensorineural hearing loss.

Negative Rinne test: if $BC > AC$ —that is, the sound in front of the ear is reported as quieter:

- ▶ identifies a significant conductive component of hearing loss of > 15 dBHL
- ▶ BUT, a **false positive Rinne** can occur if there is a severe sensorineural hearing loss in the tested ear, as the BC stimulus is heard in the non-tested ear because of transcranial transmission, and thus will be louder than AC sound. This can be overcome by masking the non-affected ear with a Barany noise box.

The Rinne test has a high specificity for conductive hearing loss, but a low sensitivity, this not reaching 90% until the air–bone gap > 30 dB

Weber's tuning fork test

The aim of Ernst Weber's test (1934) is to identify the better hearing cochlear. It is used in conjunction with Rinne's test and is of most use in patients with unilateral hearing loss:

- (1) The 512 Hz tuning fork is struck and placed to the head in the midline, either at the vertex or on the forehead
- (2) The patient is asked to say whether the sound is heard better in one ear, or equally in both ears

A central Weber is described if the tone is heard centrally

- ▶ identifies a normal hearing patient
- A lateralising Weber** is when the tone is heard to one side
- ▶ identifies the side of the better hearing cochlear
- ▶ BUT, if there is a conductive component to the hearing loss, the tone may be heard in the poorer hearing ear (see Rinne's test)

The results need to be interpreted with care and only in conjunction with further hearing tests.

overtones, two thirds of the way along its tines to minimise distortion products.

The clinical findings described above will point to the appropriate test battery to investigate further any associated hearing loss. Pure tone audiometry shows the existence and extent of hearing loss and allows determination of whether the loss is conductive, or sensorineural, or both. It requires the cooperation of the subject and as such is a subjective estimate of hearing thresholds. As a psycho-acoustic measurement, the results of audiometry may be biased by particular methods of conducting the test, and so a well defined procedure must be adopted. If the patient is unable or unwilling to cooperate, additional audiological investigation will be necessary to provide objective measures of hearing. These additional tests include measurement of otoacoustic emissions, stapedius reflex threshold measurement, and brainstem auditory evoked responses.

The purpose of aural admittance testing—that is, tympanometry—is the objective determination of middle ear pressure, the measurement of static acoustic impedance, and characterisation of the tympanometric shape. High impedance abnormalities include a perforated tympanic membrane, middle ear effusion, retracted tympanic membrane, or ossicular fixation. Low impedance abnormalities include thin atrophic tympanic membranes and ossicular disruption following head trauma.

SECTION 2

Clinical examination of eye movements

Examination of the eye movements requires optimal conditions. There should be good lighting with the patient sitting comfortably, head erect. The clinician should be aware of the visual acuity of both eyes, using the Snellen chart at 6 m. The eyes should be in the primary position of gaze and the visual target held at a distance just greater than the patient's focal point.

Cover test for strabismus

The presence of a manifest or a latent strabismus needs to be determined. Either can cause an abnormal eye movement examination because of changing optic fixation from one eye to the other. When one eye has become amblyopic, the other eye should be assessed for both clinical and electro-nystagmographic purposes. To perform the cover test, each

eye is covered in turn, either with the hand or a piece of card, close to the eye to prevent optic fixation of the covered eye. The patient is asked to fixate on a visual target and then the covered eye observed as the cover is taken away.

- (1) Manifest strabismus/exotropion is identified when the affected eye is turned outwards during normal gaze. An esotropion is identified when the eye is turned inward during normal gaze.
- (2) An exophorion in the covered eye is identified when the covered eye moves inward as the cover is removed. An esophorion is identified as the covered eye moves outwards with re-fixation.
- (3) Latent nystagmus is identified when nystagmus is seen in the uncovered eye during cover testing. Typically, the nystagmus beats away from the covered eye and is conjugate. Latent nystagmus can be uni- or bilateral and is congenitally determined. In some patients it can be associated with congenital nystagmus (vide infra).

Range of eye movements

The eyes are examined for range in both horizontal and vertical planes to a limit of 30° from the midline. The eye movements are conjugate if both eyes move together. If the movements are conjugate but the range is not full, the patient may have a gaze paresis.

Gaze paresis

A gaze paresis occurs if there is a restriction in the range of eye movements, in one or more directions. It may be nuclear or supranuclear, with reference to the oculomotor nuclei within the brainstem:

- ▶ contralateral horizontal gaze paresis occurs in lesions of the frontal eye fields in the cortex
- ▶ ipsilateral gaze paresis may identify a lesion of the tectum, or of the ponto-medullary or ponto-mesencephalic junctions
- ▶ supranuclear gaze palsy can be seen in mesencephalic lesions, in which there is loss of volitional gaze, with vertical movements being lost before horizontal. This is identified by finding a full range of eye movements in response to involuntary reflex testing—that is, vertical vestibulo-ocular reflex (VOR) testing.

Ocular paresis

Where there is a dissociation of eye movements—that is, they are dysconjugate—an ocular paresis is likely. The patient's eyes should be examined for IIIrd, IVth, and VIth nerve palsies:

- (1) The oblique muscles are tested when the eyes are adducted; the superior oblique, through its pulley system, will lower the eye and conversely the inferior oblique will elevate the eye
- (2) The rectus muscles are tested with the eye abducted, with the inferior rectus lowering the eye and the superior rectus elevating the eye.

An ocular paresis may be caused by a retro-orbital space occupying lesion or by involvement of the extra-ocular muscles—that is, thyroid eye disease, mitochondrial cytopathy or manifest strabismus. Lesions of the IIIrd, IVth, and VIth cranial nerves, or their nuclei, cause a paresis in the direction of the pull of the muscles they innervate:

- ▶ a lesion of the IIIrd cranial nerve causes the eye to be drawn down and out, and may or may not be associated with ptosis and a dilated pupil—that is, if the lesion

involves the parasympathetic fibres carried with the IIIrd nerve

- ▶ a lesion of the IVth cranial nerve causes the eye to be slightly elevated, leading to adduction of the eye and a possible head tilt to the side to the lesion, to reduce diplopia
- ▶ a lesion of the VIth cranial nerve causes a loss of abduction and again the head may compensate by rotating towards the side of the lesion.

Ocular stabilising systems

There are three visually controlled systems producing eye movements that stabilise gaze: the saccadic system, the smooth pursuit system, and the optokinetic system. The saccadic system responds to error in the direction of gaze by initiating a rapid eye movement to correct a retinal position error; the smooth pursuit system is responsible for maintaining gaze on a moving target by keeping the target within the visual field; the optokinetic system is thought to be a more primitive form of smooth pursuit, involving the whole retina instead of the fovea alone. These systems can each be tested and normal function is likely to indicate integrity of central vestibular pathways.

Saccades

This is a fast eye movement with a velocity between 350–600°/second, the velocity increasing with increased amplitude of eye movement. Saccades can be voluntary or involuntary, the former used to move the eyes between visual targets in the shortest possible time; the involuntary saccade maintains the target on the fovea when there has been slip of the retinal image, and is the fast phase of nystagmus. The saccade may be visually triggered, as in optokinetic nystagmus, or can be of vestibular or cervical origin, with a fast phase of nystagmus shifting the eyes in the direction of the ongoing head movement before the slow phase compensatory drift. Normal subjects are accurate up to a target jump of 20°, above which a small corrective saccade is required to bring the fovea on target. Overshooting is rare. The normal saccadic latency before a new saccade can be generated is 200 ms.

The ability to generate saccades depends on the integrity of projections between the frontal eye fields, the caudate nucleus, the substantia nigra reticulata, and the deep and intermediate layers of the superior colliculus. Projections are then to the para-pontine reticular formation (PPRF) and from here to the ipsilateral abducens nerve nucleus and by the median longitudinal bundle (MLB) to the contralateral oculomotor nucleus of the medial rectus. The pretectal neurons also project to the oculomotor nuclei, both sets of neurons connecting to the vestibular nuclei.

Clinical assessment

Saccadic eye movements are assessed by asking the patient to look back and forth between two targets in front of him/her, sited approximately 30° to the right and 30° to the left of the midline, respectively. Increasing the distance between the targets beyond 30° increases the chance of detecting a hypometric saccade, while reducing the distance increases the chance of detecting a hypermetric saccade.

Abnormalities of saccadic eye movements

Three variables are examined: saccadic reaction time (latency), saccadic velocity, and saccadic accuracy. Abnormalities of any of these features may be caused by

central nervous system (CNS) pathology or ocular myopathy. Peripheral vestibular pathology does not cause abnormal saccadic eye movements.

- ▶ *Internuclear ophthalmoplegia (INO)*—caused by a lesion of the ipsilateral MLB. May present as ataxic nystagmus, whereby adducting saccades ipsilateral to the lesion are slower than abducting saccades. Subtle early lesions can be revealed by electronystagmography (ENG) recordings of separate eye saccades, showing slowing of the adducting saccade on the side of the lesion with a normal velocity, but a hypermetric abducting saccade (fig 8)
- ▶ *Lesion of PPRF*—this causes loss of all types of rapid ipsilateral movements and the eyes move to the contralateral visual field. In response to vestibular or visual stimulation, the eyes display a tonic contralateral deviation with loss of the fast phase of nystagmus.
- ▶ *One-and-a-half syndrome*—seen with extensive brainstem pathology affecting both the ipsilateral PPRF and the median longitudinal bundles. This syndrome can result with a failure of conjugate gaze in one direction and an internuclear ophthalmoplegia in the other.

The pathways subserving vertical and horizontal saccades are independent, such that vertical saccades are unimpaired by lesions of the PPRF, while lesions of the mesencephalic reticular formation affect vertical saccades exclusively.

- ▶ *Cerebellar pathology*—may affect the accuracy of saccades with undershooting (hypometria) and/or overshooting (hypermetria) ipsilateral to the side of the lesion. Hypermetria is more common in cerebellar lesions than hypometria, while in intrinsic brainstem lesions, hypometria is more likely
- ▶ *Supranuclear degeneration*—MSA (Steele-Richardson-Olszewski syndrome, Shy-Drager syndrome, progressive supranuclear palsy, and Huntington's chorea), saccade reaction time is prolonged. In these conditions, hypometria cannot occur. Vertical eye movements are usually affected before horizontal, initially with upgaze more involved than downgaze and saccades more affected than pursuit

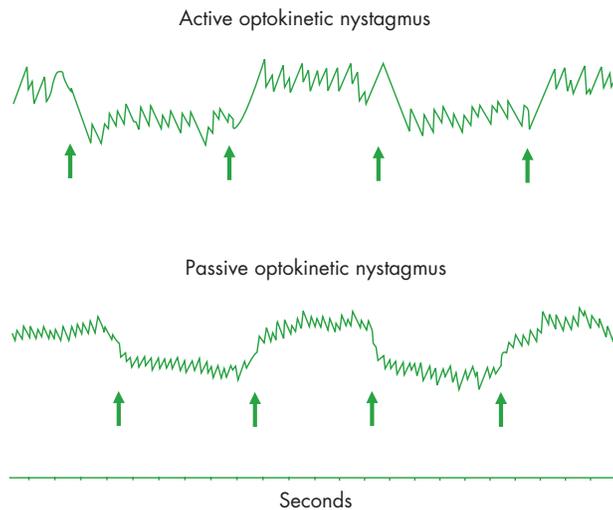


Figure 3 Active versus passive optokinetic nystagmus (OKN) (see text).



Figure 4 Optokinetic stimulus (see text). Reproduced with permission from Jaeger Toennies, Wurzburg, Germany.

- ▶ *Frontal pathology*—there may be hypometria or loss of horizontal saccades, contralateral to the side of the lesion. Vertical saccades are not affected.

Smooth pursuit

Smooth pursuit is responsible for maintaining gaze on a moving target so that the target is stabilised on the fovea. The gain of the pursuit system approaches unity at peak velocities of 30°/s or sinusoidal rotation at 0.1 Hz (Baloh *et al*²). Above a peak velocity of 60°/s, or sinusoidal rotation at 1 Hz, the gain falls off rapidly and “catch-up” saccades are observed—that is, saccadic intrusions—and the pursuit is described as broken. The smooth pursuit and the vestibular ocular reflex system are complementary in stabilising the retinal image, with the pursuit system efficient at low target velocities, and the vestibulo-ocular system efficient at high input velocities.

Clinical assessment

The smooth pursuit system can be examined clinically by moving a target—that is, the examiner's finger—slowly back and forth in a sinusoidal fashion, initially in the horizontal and then in the vertical plane, to a maximum of 30° displacement from the midline, at 0.2–0.4 Hz. In chronic peripheral vestibular disorders, smooth pursuit is normal.

Abnormalities

Impairment of smooth pursuit may be caused by lesions of the fovea, of the calcarine cortex, of the parieto-occipital cortex, the parieto-temporal region, the dorsolateral pontine nucleus, and the cerebellar-flocculus. The abnormality may be ipsilateral or bilateral. Pursuit eye movements are symmetrically affected by age, psychotropic medication, alcohol, anticonvulsants, and vestibular and CNS sedatives.

Optokinetic nystagmus

The function of optokinetic nystagmus (OKN) is thought to be the stabilisation of the eyes relative to space during slow head movements in the low frequency range, ill served by the VOR—for example, the person looking at the scenery from a moving vehicle. The optokinetic system includes the peripheral retina, the accessory optic tract, the vestibular nuclei, and the reticular formation. There are two types of OKN (fig 3):

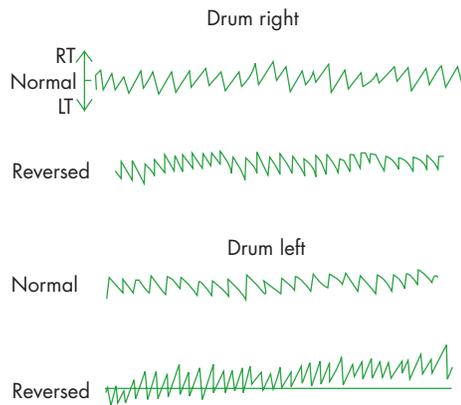


Figure 5 Congenital nystagmus showing the optokinetic response in a patient with congenital nystagmus—that is, reversal of OKN with the slow phase of the patient's eyes in the opposite direction to the direction of the OKN drum.

- ▶ *Cortical optokinetic nystagmus*—active or “look” OKN, induced more through foveal input, and can be induced both by using a small drum and by a full field optokinetic stimulus
- ▶ *Subcortical optokinetic nystagmus*—passive or “stare” OKN, in which it is believed that the peripheral retina is involved without participation of the cerebral cortex

Although the two tracings appear similar, in the active situation, the slow component velocity is similar to the speed of the drum—that is, the gain approaches 1. Also, with reversal of the drum direction, the eyes deviate in the direction of the slow component—that is, the same as the direction of movement of the drum. However, in the stare condition, subserved by the subcortical pathway, the slow component velocity is consistently less than drum velocity, and with drum reversal, the direction of eye deviation is in the direction of the fast phase of the nystagmus.

Clinical assessment

A qualitative assessment may be undertaken at the bedside or in the outpatient department using a small handheld or mechanically driven optokinetic drum. This is a cylinder 30 cm diameter that can be rotated to elicit nystagmus either in the horizontal or vertical plane, at speeds from 40°/s upwards.

For quantitative purposes, more precise stimulus parameters are obtained by seating the patient inside a large, striped, rotating drum and stimulating the entire visual field (fig 4).

Clinical abnormalities

- ▶ *Peripheral lesions*—the imbalance of vestibular tone resulting from lesions of the labyrinth and VIIIth nerve can give rise to a directional preponderance with the hand held drum. It is best seen with direct observation of the eyes with repeated abrupt reversal of the drum direction. These abnormalities are rarely seen with full field OKN
- ▶ *Central lesions*—abnormalities of OKN tend to mirror abnormalities of smooth pursuit, and abnormalities of fast components mirror abnormalities of voluntary saccades. Lateralised lesions of the parieto-occipital region, brainstem and cerebellum result in impaired OKN when the stimulus is moved toward the damaged side.

- ▶ *Congenital nystagmus*: reversal of OKN is a significant feature in many cases of congenital nystagmus (fig 5). The nystagmus beats paradoxically in the opposite direction to that anticipated from the direction of drum movement.

Assessment of nystagmus

Nystagmus is a combination of alternating slow and fast phase eye movements in opposite directions. For clinical purposes the direction of nystagmus is defined by the fast phase. It can be physiological or pathological and pathological nystagmus can be congenital or acquired. In principle, large amplitude nystagmus should be considered as central in origin, and is only likely to be peripheral if seen in the first few days of vestibular neuritis or an acute episode of Menière's disease.

Physiological vestibular nystagmus

The semicircular canal–ocular reflexes produce eye movements that compensate for head rotations. With small amplitudes of head displacement, there is a slow compensatory eye movement in the direction opposite to rotation, serving to stabilise the gaze. With a larger stimulus the slow vestibular induced eye movement deviation is interrupted by a fast eye movement in the opposite direction, generating physiological nystagmus. This type of nystagmus can be induced both by rotary chair testing and caloric irrigations but also by extremes of eye deviation—that is, more than 30° laterally from the primary position (physiological end point nystagmus).

There is a relation between the magnitude of nystagmus and the state of arousal in human subjects during vestibular stimulation. A subject who is allowed to daydream has a lower slow component velocity than the subject who is asked to perform continuous mental arithmetic. Subjects experiencing repeated angular accelerations—for example, ice skaters and dancers—may display permanent habituation of the response with a reduction or loss of nystagmus in response to vestibular stimulation.

Pathological vestibular nystagmus (spontaneous nystagmus)

Spontaneous nystagmus results from an imbalance of tonic signals arriving at the oculomotor neurones. Because the vestibular system is the main source of oculomotor tonus, it is the driving force of most types of spontaneous nystagmus, hence the name vestibular nystagmus. There is a constant

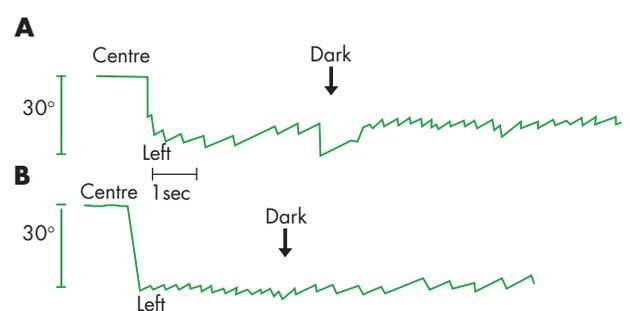


Figure 6 Peripheral versus central nystagmus identifying the effects of gaze testing in a patient with: (A) a peripheral vestibular deficit—the spontaneous nystagmus on gaze left is enhanced in the absence of optic fixation; (B) a gaze evoked nystagmus—the nystagmus decreases in frequency and slow component velocity in the absence of optic fixation.



Figure 7 The Dix-Hallpike manoeuvre (see text).

drift of the eyes towards the side of the lesion, interrupted by a fast component in the opposite direction. The lesion may occur in the labyrinth, the vestibular nerve, or in the vestibular nucleus.

Spontaneous nystagmus is invaluable in siting vestibular and neurological disease. For full assessment, the nystagmic response produced by (1) change of eye position, and (2) the presence or absence of optic fixation, needs to be documented. Nystagmus is graded using Alexander's law: if it is only present when the eye is deviated towards the fast phase it is 1°; if it is also seen in the primary position it is 2°; and if it is also seen with the eye deviated towards the slow phase it is 3°. The vestibular lesion may only be detected with the removal of optic fixation if the lesion is small or compensation at a central level has occurred. This is an important criterion for identifying nystagmus caused by peripheral pathology—that is, the nystagmus displays an increase of amplitude with the removal of optic fixation and thus the nystagmus may be detected in the dark by ENG or video-oculography.

Gaze evoked nystagmus

Patients with gaze evoked nystagmus are unable to maintain stable conjugate eye deviation away from the primary position. The eyes drift backwards towards the centre with an exponentially decreasing waveform. Corrective saccades constantly reset the eye in the desired position, thus gaze evoked nystagmus is always in the direction of gaze. In the absence of optic fixation, the frequency and slow component of velocity decrease (fig 6). Dysfunction may be secondary to a lesion anywhere from the multiple brain centres controlling conjugate gaze to the neuromuscular junction. It is not

caused by a peripheral vestibular lesion. Symmetrical gaze evoked nystagmus is commonly observed following the use of anticonvulsants, particularly phenytoin and phenobarbitone, and ingestion of psychotropic drugs and alcohol. Asymmetrical horizontal gaze evoked nystagmus is likely to indicate a structural brain lesion.

Positional nystagmus

Nystagmus can be elicited by critical head positioning in certain pathological states. The Dix-Hallpike head manoeuvre (fig 7) is a valuable test in such patients and can distinguish between the peripheral nystagmus of benign paroxysmal positional vertigo (BPPV), central positional nystagmus, and atypical positioning nystagmus. The test consists of:

- ▶ seating the patient appropriately on a couch so that when supine, their head will extend over the end of the couch
- ▶ the patient is asked to remove their spectacles and is warned they may feel dizzy as a result of the test
- ▶ he is asked not to close his eyes but to keep his gaze centred on the examiner's forehead
- ▶ the patient's head is turned 30–45° towards the examiner, and moved rapidly into the lying position with the head hanging 30° over the back of the couch (in this way the posterior semicircular canal of the undermost ear is moved directly through its plane of orientation)
- ▶ the patients' eyes are observed for nystagmus for up to one minute
- ▶ the manoeuvre is repeated for both the right and the left ear undermost positions.

Peripheral positional nystagmus (caused by BPPV)

BPPV was first described by Barany in 1921 and is diagnosed using the Dix-Hallpike manoeuvre. With the pathological ear under most, the Hallpike test produces the following classical signs

- ▶ Latent period to onset of nystagmus from 2–45 seconds
- ▶ Development of torsional nystagmus with the fast phase towards the ground (geotropic)
- ▶ Associated vertigo and autonomic symptoms
- ▶ Adaptation of vertigo and nystagmus on maintaining the head hanging position
- ▶ Reversal of nystagmus on returning to the upright position
- ▶ Fatiguing of symptoms and signs on repeating the test.

Central positional nystagmus

Typically, there is no latency to the onset of nystagmus, frequently no vertigo, and no adaptation of fatigability (table 1). The direction of nystagmus may be towards the uppermost ear, or may be vertical in direction. Multiple sclerosis, Arnold-Chiari malformation, and cerebellar vascular disease may produce positional nystagmus that may be the only sign of posterior pathology.

Congenital nystagmus

The patient with congenital nystagmus rarely complains of oscillopsia, but has a central eye movement abnormality. The nystagmus is in the horizontal plane and may change direction. There is a null point, which is often the head position the patient adopts for reading; the slow phase is dysmorphic and may be exponential as demonstrated on the ENG, and characteristically there is reversal of OKN—that is, the slow phase of OKN does not match the direction of drum rotation.

Table 1 Comparison of benign paroxysmal positional nystagmus (BPPN) and central positional nystagmus (CPN)

	BPPN	CPN
Latent period	2–45 seconds	0 seconds
Adaptation	Within 30 seconds	Persisting
Fatigability	Disappears on repetition	Persists
Vertigo	Present, sometimes severe	Usually absent, or very mild
Direction of nystagmus	Torsional and geotropic	Any
Incidence	Common	Rare

Periodic alternating nystagmus

Periodic alternating nystagmus changes direction with a change of head or eye position. Cycle length varies from 1–6 minutes with null periods of 2–20 seconds. The precise site of the lesion is known, but both the cerebellum and the caudal brainstem have been implicated.

Torsional nystagmus

This nystagmus is a rotation of the eye, beating around the visual axis. The direction of beat is better specified in right-left terms to avoid confusion—that is, “clockwise” nystagmus is as seen from the examiner’s point of view. It is usually caused by a lesion in the area of the vestibular nuclei on the opposite side to the beat direction as in Wallenberg’s syndrome.

Pendular nystagmus

This is seen in association with longstanding or congenital visual defects (when it is not associated with balance symptoms) or it can develop weeks to months after structural brainstem disease. In the latter, patients usually show cerebellar or pyramidal features and describe oscillopsia which is largely unchanged by head movements.

Monocular nystagmus by definition only occurs in one eye and has been reported in a number of ophthalmological and neurological conditions.

Tests of stance and gait

Romberg test

The Romberg test was described in 1846 in patients with dorsal column loss as a result of tabes dorsalis. The test is described as positive when there is increased body sway with the eyes closed when the patient is standing with his feet close together. The principle lying behind this test is that balance is maintained with minimal physiological sway when all three sensory inputs are functioning—that is, vision, vestibular input, and proprioceptive input. With the loss of one or more of these inputs, there will be increased physiological sway. Unsteadiness on Romberg testing can also occur with acute vestibular deficits and with cerebellar disease, although in the latter, the effects of eye closure theoretically should not affect sway.

Unterberger’s test

Unterberger described the tendency of vestibular stimulation to turn the patient in the earth’s vertical axis when walking. His test identifies that the direction of turning in patients with unilateral vestibular deficits coincides with the direction of past-pointing and falling—that is, in the direction of the slow component of nystagmus. The test is performed by asking the patient to stand with their arms extended and thumbs raised, and then to close their eyes. The patient is asked to march on the spot for about 50 steps and the angle of rotation as well as forwards and backwards movements is recorded. There is, however, a pronounced variability in the rotation angle from one subject to another and in the same subject on repeated testing, and the outcome should only be used in the context of the rest of the vestibular test battery.

Gait test

This is a 5 m walk, firstly with the eyes open and then with the eyes closed, with the patient walking at normal speed towards a fixed target and the examiner close to one side for safety reasons. As with the Unterberger’s test, patients with recent unilateral vestibular lesions tend to deviate

towards the side of the lesion. This test is fraught with false negatives.

Tandem gait test

These tests are useful for assessing vestibulospinal function. When performed with the eyes open, tandem walking is primarily a test of cerebellar function because vision compensates for chronic vestibular and proprioceptive deficits. Tandem walking with the eyes closed provides a better test of vestibular function as long as cerebellar and proprioceptive functions are intact. The subject is asked to start with feet in the tandem position and arms folded against the chest and to make 10 steps at a comfortable speed. Most normal subjects can make a minimum of 10 accurate tandem steps in three trials.

SECTION 3

Vestibular testing

Electronystagmography (ENG)

Although a good examination of eye movements can be made by direct observation, recording techniques including ENG, video-oculography, and spiral coil recordings allow a more detailed evaluation and provide a permanent record for comparative purposes.

ENG is the simplest and most readily available system for recording eye movements. An electrode placed laterally to the eye becomes more positive when the eye rotates towards it and more negative when it rotates away. The voltage change represents the change in eye position as only small angular movements are involved in nystagmus and the relation between voltage change and eye movement is virtually linear at the small degrees of arc. The polarity of the recording is arranged so that a deflection of the eye to the left causes a downward deflection of the pen and a deflection of the eye to the right causes an upwards deflection. The sensitivity of the ENG can consistently record eye rotations of 0.5°. This

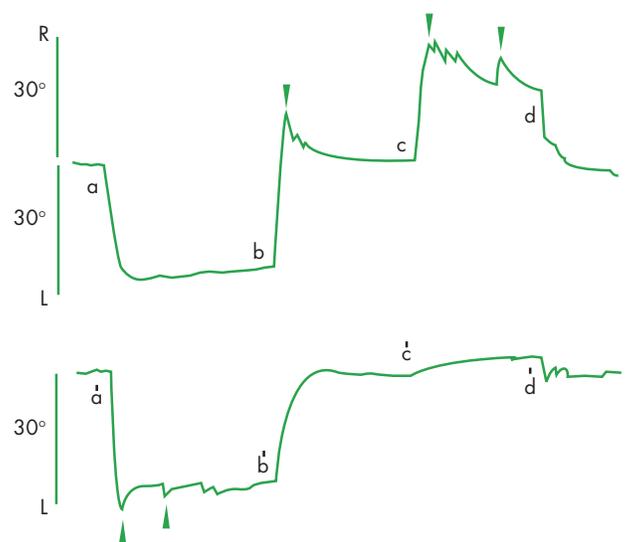


Figure 8 Internuclear ophthalmoplegia (INO). The upper trace shows the movements of the right eye and the lower trace shows those of the left. At point a/a' the eyes are looking straight ahead; at b/b' the eyes are looking out at 30° to the left; at c/c' the eyes are again looking straight ahead; at d/d' the eyes are looking out at 30° to the right. The trace identifies a left INO with failure of the left eye to adduct (d') and nystagmus in the abducting right eye (d).

Standard protocol for ENG testing with rotary chair

- ▶ Calibration—eyes centred, eyes right at 30°, eyes re-centred, eyes left 30 (repeat)
- ▶ Gaze testing to characterise nystagmus:
 - (1) centre gaze, eyes open, then in dark for 20 s (that is, optic fixation inhibited)
 - (2) gaze held at 30° to the right for 10 s, then 20 s in the dark
 - (3) gaze held at 30° to the left for 10 s, then 20 s in the dark
 - (4) if vertical nystagmus identified—vertical eye movement recordings required with: (i) gaze upwards at 30° for 10 s, then 20 s in the dark; (ii) gaze down 30° for 10 s, then 20 s in the dark
- ▶ Visual stimuli (*using laser target for 1, 2 and 3)
 - (1) Saccades—between two targets at 5–30 apart
 - (2) Separate eye saccades—recordings taken from each of the two eyes, paper speed 10× normal, then follow protocol for gaze testing as above
 - (3) Smooth pursuit—at 0.1, 0.2, 0.3, 0.4 Hz, maximum velocity 40°/s
 - (4) OKN—full field, constant velocity around vertical central axis, at 40°/s, reverse direction after 9 s
- ▶ Rotary chair testing
 - (1) Impulsive rotational testing—acceleration rise time of < 1 s to a constant velocity of 60°/s
 - (2) Sinusoidal VOR testing—at 0.1, 0.2, 0.3, 0.4 Hz, maximum velocity 40°/s
 - (3) VOR suppression—at 0.1, 0.2, 0.3, 0.4 Hz, maximum velocity 40°/s

*The advantage of a laser target is that its size remains constant with distance from its source, unlike conventional light sources, and an infinite number of computerised projection paradigms are possible for different test and research purposes

sensitivity is less than that of direct visual inspection which is approximately 0.1°.

The plane of the recording electrodes defines the plane of recorded eye movement—that is, electrodes attached medially and laterally to the eye will record the horizontal components of eye movement, whereas those attached above and below will record vertical components. A single channel ENG machine summates the horizontal movements of both eyes from bitemporal recordings onto the same trace. The two channel ENG machine can record the movement of each eye separately (fig 8), and with four channel use the technique can simultaneously record the vertical movements of each eye as well as the horizontal movements. The paper speed of 10 mm/s is used although when saccadic accuracy and velocity are being tested, 100 mm/s or even faster may be necessary. Calibration is performed, so that a standard angle of eye deviation is represented by a known amplitude of pen deflection. Commonly the calibration is adjusted to obtain 10 mm of pen deflection per 10°.

Clinical relevance of ENG

The main advantage of ENG recordings is that some patients demonstrate nystagmus that is only identifiable when optic fixation is removed.

- ▶ *Peripheral vestibular disorders*—unless acute, these deficits are unlikely to be associated with nystagmus in the presence of optic fixation but can reveal nystagmus of

increased amplitude in darkness. The nystagmus is unidirectional with the largest amplitude on horizontal gaze towards the direction of the fast component

- ▶ *Vestibular nuclei lesions*—in darkness the amplitude of the nystagmus may hardly alter but the velocity of the slow phase may be decreased. Often the nystagmus is bidirectional
- ▶ *Cerebellar lesions*—these may be associated with pathological square waves with duration of less than 200 ms. With direct current ENG recordings a characteristic abnormality of cerebellar pathology is the failure to maintain lateral gaze in darkness with a slow drifting movement of the eyes. Rebound nystagmus can also be seen, but when cerebellar, it is transitory and will persist for a maximum of 20 seconds. Patients with cerebellar disease may also have difficulties in executing commands of saccadic movements. When asked to turn their gaze laterally quickly they overshoot the target.

Rotary chair testing allows measurement of the eye movement response to precise vestibular stimuli and can be of immense clinical value. The VOR (vestibulo-ocular reflex) provides a simple example of a reflex arc comprising the vestibular sense organ, the primary, secondary, and tertiary vestibular neurones and the effector organ, the oculomotor muscle. Angular acceleration in the plane of the semicircular canal leads to endolymph displacement in a direction opposite to that of rotation and in consequence the cupula of that canal deviates in the same direction as the endolymph, resulting in a change of vestibular tonus, an excitatory stimulus being matched by an inhibitory stimulus from the opposite side. As a result there is an impact on the pair of muscles producing the compensatory eye movement—that is, excitation of the antagonist muscle and disinhibition of the agonist muscle. An acceleration to the right in the plane of the horizontal canal will produce deviation of the eyes to the left.

VORs act during all natural head movements in life with coordination with visual and cervico-ocular reflexes to provide the most appropriate eye position and eye stability during head movements. Vision has a powerful suppressive effect on vestibular nystagmus that can, however, be seen using Frenzel's glasses or VOG.

Types of rotary chair stimuli

- ▶ *Impulsive (step velocity) stimuli*—Constant velocities such as 40, 60, 80 or 120°/s are attained with an abrupt acceleration of the chair, brought to constant velocity within 1 second. This constant velocity is maintained for up to 2 minutes while the nystagmic response dies away. The chair is suddenly brought to rest with the same deceleration and the normal limits of nystagmus intensity are established with normal subjects. It provides a rapid assessment of gain (peak slow component velocity ÷ change in chair velocity) and the time constant (time for the slow component velocity to fall to 37% of its initial value) of the canal reflex.
- ▶ *Sinusoidal stimuli*—To and fro swinging movements of the chair around its vertical axis are programmed with variable stimulus parameters—that is, frequency and amplitude. The threshold for recordable nystagmus, defined as the angular acceleration maintained for 20 seconds that will produce nystagmus, is 0.15°/s² in the absence of optic fixation. With optic fixation the nystagmus threshold is raised and is normally about 1°/s². With

computer software analysis, sinusoidal VOR at different frequencies can be used to calculate gain and phase of the peripheral vestibular response.

Rotational testing is normally performed in darkness but sinusoidal rotation can also be performed with the eyes focused on a target that revolves with the patient, around the vertical axis. This has the purpose of allowing VOR suppression to be assessed. This is an important test of central vestibular function, as visual suppression of the VOR is mediated by central vestibular pathways.

Clinical relevance of rotary chair testing

Rotational stimuli can be used to demonstrate a directional preponderance as determined by the ratio of the duration of nystagmus following the onset of acceleration to that following deceleration. A disadvantage is that both labyrinths are tested simultaneously and a unilateral dysfunction may be difficult to identify if the lesion is old and the patient is well compensated. It is of particular value in the following situations:

- ▶ A negative caloric test, where high frequency oscillation/high intensity acceleration may give evidence of some residual vestibular function
- ▶ Investigation of visuo-vestibular interactions: failure to suppress the vestibulo-ocular reflex with fixation is evidence of central vestibular dysfunction.

With computerised analysis of responses to rotary chair testing, results can be depicted in a quadrantic fashion following the sequence of start/stop stimuli in a clockwise then anticlockwise direction. These displays include a mathematical computation of directional preponderance using slow phase velocity and/or durational criteria (vide infra). Dimitri *et al*³ applied multivariate classification techniques of sinusoidal harmonic accelerations and measured asymptotic gain and the time constant, and were able to demonstrate a minimal misclassification rate of 3.4% when comparing 57 normal with 30 patients with peripheral vestibular deficits—that is, as defined by a total canal paresis on caloric testing.

Video-oculography

This is a technique for observing eye movements whereby an infra-red camera is mounted within goggles and connected to a video monitor. Observation and recordings of the eye movements in the absence of optic fixation can be made in response to a variety of stimuli and this technique is now a standard investigative tool in many audiology departments. A test protocol can be performed which allows videomonitoring of the following:

- ▶ spontaneous nystagmus
- ▶ head shaking nystagmus
- ▶ passive head tilt
- ▶ rotation of 180°
- ▶ body rotation with respect to head.

As a technique for bedside vestibular testing, Vitte *et al*⁴ clearly demonstrated the value of VOG for identifying and classifying peripheral vestibular lesions.

Caloric testing

This is the most widely available of all the vestibular tests, and for many otologists is the cornerstone of vestibular diagnosis. Its great value is that it allows each labyrinth to be tested separately. The stimulus is easy to apply and involves

inexpensive methodology. The test remains unrivalled as a method of demonstrating a peripheral vestibular deficit.

Principles of caloric testing

After irrigation of the ear with water 7°C below (30°C), and then 7°C above (44°C) body temperature, a gradient is set up between the EAM and the two limbs of the horizontal canal. This is by virtue of the position of the patient, who is reclined on the couch and whose head is at 30° to the horizontal. This means that the horizontal semicircular canal becomes vertical and the temperature gradient crosses from one side of the canal to the other. It is believed that the endolymph circulates because of the difference in the specific gravity on the two sides of the canal. With warm water, there is ampullo-petal flow, with cupular deflection towards the utricle, resulting in activation of the VOR, a sensation of vertigo and horizontal nystagmus directed towards the stimulated ear. There are some questions regarding this convection theory, because caloric nystagmus still occurs in space under micro-gravity conditions.

The Hallpike-Fitzgerald bithermal caloric test has been available to clinicians for more than 60 years. Each ear is irrigated in turn for 40 seconds with first 30°C, and then 44°C water. Inspection of the tympanic membrane after warm irrigation confirms an adequate stimulus if a red flush is seen on the tympanic membrane.

Direct observation of the eyes allows the end point of the nystagmic reaction to be measured. During the procedure, the patient is asked to direct his gaze on a fixation point on the ceiling above his head, making the end point easier to determine. At this point the lights are switched off and the eyes observed with Frenzel glasses or infra red gun and under normal situations the vestibular nystagmus would be expected to reappear. The end points of each test are graphically recorded.

Quantitative analysis

Normally, nystagmus ceases between 90–140 seconds after onset of irrigation and will return for up to a further 60 seconds after the removal of optic fixation. Two characteristic patterns of response may appear, either separately or in combination.

- (1) A total canal paresis is the complete loss of labyrinthine function in one ear. This is seen when there is a total absence of nystagmus following both 30°C and 44°C irrigations, even in the absence of optic fixation. Ideally, the test should be repeated using cold water at 20° for 60 seconds to confirm the result. It may reflect an ipsilateral lesion of the labyrinth, the VIIIth nerve, or the vestibular nuclei within the brainstem.
- (2) A directional preponderance occurs when the responses to thermal irrigations produce an excess of nystagmus in one direction—that is, towards either the right or the left. It indicates an imbalance of vestibular tone arriving at the oculomotor nuclei and may result from peripheral vestibular lesions (that is, the labyrinth, the VIIIth nerve or the nuclei) or from central vestibular lesions (that is, within the cerebellum or brainstem). With more pronounced degrees of vestibular tone imbalance, spontaneous nystagmus makes its appearance.

Figures for duration of nystagmic responses in seconds can be entered into the Jongkees formula, which allows

calculation of a percentage figure expressing the degree of canal paresis or directional preponderance:

$$\text{Canal paresis (\%)} = \frac{(R30^\circ + R44^\circ) - (L30^\circ + L44^\circ)}{(R30^\circ + R44^\circ + L30^\circ + L44^\circ)} \times 100$$

$$\text{Directional preponderance (\%)} = \frac{(L30^\circ + R44^\circ) - (L44^\circ + R30^\circ)}{(L30^\circ + R44^\circ + L44^\circ + R30^\circ)} \times 100$$

The optic fixation index (OFI) is calculated by dividing the summed durations in the light by the summed durations in the dark. If there is no enhancement in the absence of optic fixation—that is, the OFI is 1.0—the cause may be central—that is, in the cerebellum or the vestibulo-cerebellar tracts. Bilateral decreased caloric responses may indicate bilateral vestibular impairment, or may be the result of vestibular habituation—for example, in professional acrobats, ice skaters, and ballet dancers (OFI < 0.5).

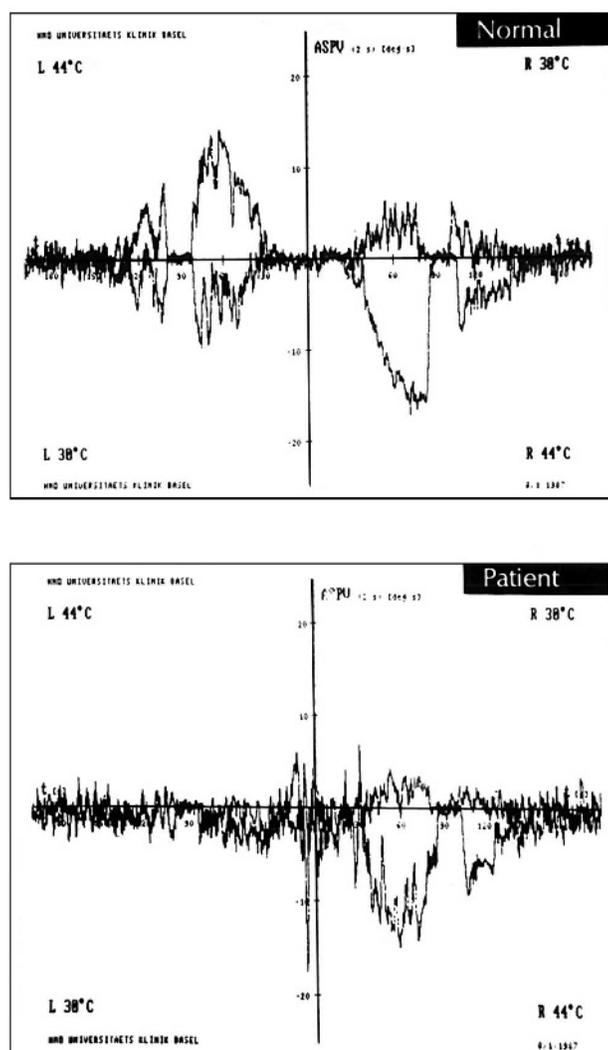


Figure 9 Four quadrant display of caloric irrigations showing a graphic record of a set of normal responses to caloric irrigation in the upper four quadrant display, with the lower four quadrant display showing a partial canal paresis on the left. From Toennies Computer-Assisted Diagnostic Systems, with permission of Professor John Allum, Basel, Switzerland, and Jaeger Toennies, Wurzburg, Germany.

ENG caloric recordings

There are both advantages and disadvantages associated with the use of ENG caloric testing. It provides a permanent record of the caloric response in both light and dark, and allows individual features of the nystagmus—that is, slow component velocity, inter-beat frequency, and the amplitude—to be analysed and a permanent record kept (fig 9). The comparative disadvantages when compared with the Fitzgerald-Hallpike technique are that:

- ▶ it is difficult to detect the end point of the nystagmus as well as can be done with the naked eye
- ▶ recording the caloric nystagmus with the eyes closed is compromised by Bell's phenomenon
- ▶ other nystagmic components in the vertical direction are missed—that is, torsional nystagmus—which may give additional diagnostic information.

A direct comparison has been made between the maximum slow component velocity and the durations of the four caloric responses in 25 normal subjects. The durations were relatively stable as a parameter, whereas the slow component velocities showed considerable variations in some subjects. Further analysis showed that the test/re-test unreliability of the slow component velocity was unacceptably high. Direct visual observation has the advantage that the end point of the nystagmus can be estimated more reliably both with and without optic fixation, but does rely on an experienced observer.

Closed circuit and air caloric testing

Some commercial systems allow warming and cooling of the EAM with closed irrigation systems and alternatively with the use of air. The problem with the former is that the tube in which the water is flowing does not fully occupy the EAM, thus reducing the effectiveness of the stimulus; and with the latter, the specific heat of air is much lower than that of water, which means a greater temperature differential is required to effect the same temperature gradient across the labyrinth—that is, a hot air stimulus of 50°C which needs to be delivered for 60 seconds will give an equivalent to the 44°C water stimulus, but may be poorly tolerated by patients.

Caloric testing is an essential part of the evaluation of the dizzy patient. It tests both labyrinths individually and does not require sophisticated instrumentation if water irrigations are to be used. The quantification and normal values of the measured parameters of the caloric induced nystagmus have been well established. The Jongkees formula is a validated measure to calculate both canal paresis and directional preponderance.

Posturography

It is well established that alterations in vestibular function may profoundly affect posture. Postural control is a vital physiological function if we are to continue any of our daily activities, and is determined by a complex sensory motor feedback system dependent on a variety of coordinated reflexes. Only in the last couple of decades have objective measures of vestibulo-spinal postural reflexes been possible.

Clinically the Romberg's test has been used to assess postural stability. During normal standing, the body is in continuous motion—that is, physiological sway—even when attempting to remain still. This is an active process, whereby any loss of balance is compensated by movement of the



Figure 10 The Equitest balance platform allowing sway referencing of both the support surface and the visual surround. Reproduced with permission from NeuroCom International Inc, USA.

body's centre of gravity. These movements result in visually detectable sway movements that maintain the body's centre of gravity vertically over the base of support.

Static force plate posturography was an early method of measuring body sway. A typical force plate consists of a flat rigid surface supported on three or more points by independent force measuring devices. As the patient stands on the fourth plate, the vertical forces recorded by the measuring devices are used to calculate the position of the centre of the vertical forces exerted on the force plate over time. With the height and weight of the patient, a computer model of body dynamics can be used to derive the centre of gravity sway angle over time.

Moving platform posturography has been designed to overcome the limitations of the static force platform by controlling the relative contributions of visual, somatosensory, and vestibular input. Commercially available platforms calculate body sway based on changes in horizontally (sheer) and vertically oriented (torque) strain gauges mounted under the support surface. The Equitest platform (fig 10) utilises the concept of sway referencing—that is, both the support surface and the visual surround can be modulated in phase with the patient's own body sway. This coupling of either the platform or visual surround to the sway of the subject allows:

- ▶ the angle between the foot and the lower leg to be maintained at a constant value (minimising somatosensory input)

- ▶ the visual input to remain constant despite the subject's sway.

Six conditions are used in the sensory organisation testing and analysis of the sway scores allows each of the three principle balance sensors to be isolated and comparisons made to assess the sensory preference of the individual.

With the moving platform, posturography has been shown to be of great benefit in rehabilitation but limited as a diagnostic test with a sensitivity of only 50%.⁵ The sensory organisation test battery identifies abnormal patterns of postural control, not adequately assessed by studies of the vestibulo-ocular reflex alone, and examines the utilisation and integration of visual and somatosensory input with vestibular information, especially under conditions of sensory conflict.

Limitations of vestibular tests

A major disadvantage of all types of vestibular test is that normative data are not universal and therefore each dataset needs to be collected for each laboratory before it can be used for clinical diagnosis. The equipment for the rotary chair and posturography is expensive and requires space and dark rooms. There is no gold standard for vestibular testing and the results of all the tests in the battery must be used in conjunction to develop the diagnosis.

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