Visual, Auditory, and Vestibular Systems (Dr. Merchut)

Visual System

1. Bedside examination and related abnormalities

The basic bedside examination of the visual system involves measurement of visual acuity, evaluation of the visual fields, and the ophthalmoscopic viewing of the optic disc and adjacent blood vessels and retina. **Visual acuity** pertains to the smallest row of numbers or letters a patient can accurately read, which is a function of central vision involving the macula. (Color vision is also a macular function.) Each eye is tested separately by having the patient read the smallest numbers or letters possible on a Snellen wall chart several feet away. **Visual acuity is recorded as a fractional number, which compares the patient's vision with the normal population**. For example, if the patient can read nothing smaller than the 20/100 line on the chart, this acuity of 20/100 means that what he or she can read at a distance of 20 feet, the normal population can read at a distance of 100 feet. Normal is thus 20/20. A pocket visual acuity chart may be used as well here, but reading it several inches away is problematic for older patients with presbyopia, or age-related impairment of near vision. An abnormal visual acuity may be due to an ocular problem, where the patient "needs glasses." This acuity problem is normalized with corrective lenses. Improved acuity when looking through a pinhole in a plastic card also suggests the problem is ocular. **The impaired visual acuity from a lesion of the optic nerve or macula, however, is not improved when looking through a pinhole, or by use of corrective lenses.**

The **visual field** examination mainly tests peripheral vision, which is "less sharp or acute" than central vision. Visual field deficits are traditionally recorded on circular or polar graphs "as the patient sees them." A circular graph on the left represents the visual fields of the left eye, while a circular graph on the right represents the visual fields of the right eye. The right side of each circle would be blackened if the patient had trouble seeing to the right. **Perimetry** is a more quantitative method of visual field testing, where a computer randomly flashes "dots" of light on a test screen, which the patient acknowledges by pushing a button. The bedside method of visual field testing is less precise, but with practice reliably detects clinically significant deficits. Visual fields are tested "by confrontation" with patient and examiner facing and "focused on" each other, "eye to eye," allowing the examiner to see whether a visual target (often the examiner's fingers) visible in the examiner's visual field is likewise perceived in the patient's visual field. The patient covers one eye at a time, and is asked to point out or acknowledge a subtle flicker of the examiner's finger, or the number of fingers shown, in alternating quadrants of the visual fields (Fig. 1).

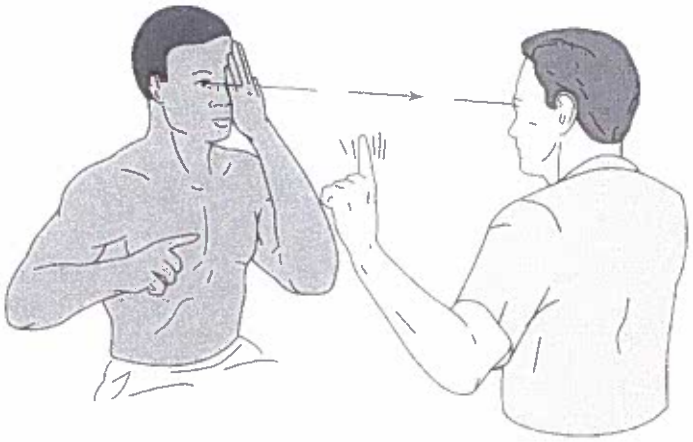

Fig. 1 Confrontation testing of visual fields.

Everyone normally has a physiological "blind spot" in the temporal visual field of each eye, since the optic disc (optic nerve head) in the nasal retina is devoid of rods and cones. This normal blind spot is more easily mapped out by perimetry testing of the visual fields, and abnormally enlarges in size in the presence of **papilledema**, where increased intracranial pressure causes the optic nerve to appear swollen on ophthalmoscopic examination. **Scotomas** are defined as pathological, abnormal blind spots elsewhere in the visual field of one eye, where vision is lost or decreased. The patient may report this deficit ("Looking directly at you doctor, I see your whole face clearly except for your left ear.") or it may be detectable only when visual fields are tested by perimetry or by confrontation. Scotomas are **due to lesions in the retina** (infections or inflammation, macular degeneration, or retinal detachments) **or optic nerve** (demyelination or ischemia). Patients with abnormally narrowed or smaller visual fields, referred to as "constricted or contracted" visual fields, may have underlying glaucoma or a retinal degenerative disease. As the examiner moves farther away from such a patient, even these "constricted" visual fields should enlarge in cone-shaped fashion, when examined again at a distance. "Tunnel vision" refers to the same narrow breadth of visual fields whether tested close to, or farther from, the patient and usually reflects a psychiatric rather than a neurological problem.

Before exploring more complex visual field deficits, it may be helpful to think of the optical elements of the eye working as a camera. A visual image in the right side of the patient's world is detected by the left halves of each retina (nasal right retina and temporal left retina), and are transmitted by optic nerve fibers which, beyond the optic chiasm, are continued as a pathway through the left optic tract, then the left optic radiations, ending in the left occipital visual cortex. Post-chiasmal lesions in these left brain pathways thereby cause visual impairments in the right side of the patient's world (D, E, F, and G in Fig. 2). A visual image in the uppermost part of the patient's world is detected by the inferior portions of the retinae, and these inferior retinal fibers continue past the optic chiasm as the inferior optic radiations of the temporal lobe. A temporal lobe lesion involving these inferior optic radiations produces a contralateral deficit which is in the patient's superior visual world (E in Fig. 2).

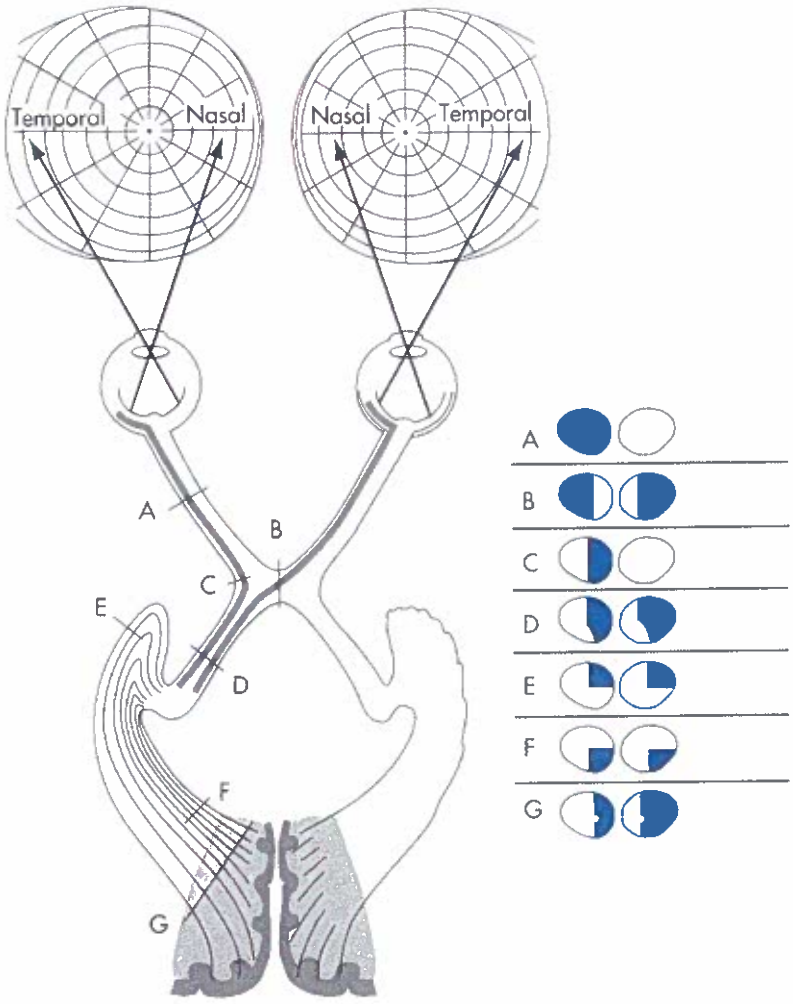

Fig. 2

Visual field defects. Correlation of various lesions with types of visual loss. (Modified from Homan J: *A textbook of surgery*, ed 6, Springfield, Ill., 1945, Charles C Thomas.)

Defect	Lesion
(A) Blindness of left eye	Left optic nerve (if ocular/retinal causes excluded)
(B) Bitemporal heteronymous hemianopsia	Inner optic chiasm
(C) Binasal heteronymous hemianopsia	Outer optic chiasm (shown only on left)
(D) Right homonymous hemianopsia	Left optic tract
(E) Right superior homonymous quadrantanopsia	Left inferior optic radiation (temporal lobe, Meyer's loop)
(F) Right inferior homonymous quadrantanopsia	Left superior optic radiation (parietal lobe)
(G) Right homonymous hemianopsia (with macular sparing)	Left occipital lobe

Hemianopsia describes visual loss or impairment in half (nasal half or temporal half) of the visual field of each eye (Fig. 2). A **homonymous hemianopsia is a deficit of the nasal half of one eye and the temporal half of the other eye**. Such homonymous visual losses affect the right side (nasal half of left eye, temporal half of right eye) or left side (nasal half of right eye, temporal half of left eye) of the patient's visual world. A homonymous hemianopsia is produced by a lesion in the contralateral **optic tract** (D in Fig. 2), or the contralateral inferior (temporal lobe) and superior (parietal lobe) **optic radiations** (E plus F in Fig. 2), or the contralateral **occipital lobe** (G in Fig. 2). Congruent nasal and temporal visual field deficits are almost identical if superimposed, and **congruence increases when the lesion is more posterior** (occipital lobe or superior and inferior optic radiations) than anterior (optic tract). A homonymous hemianopsia with **macular sparing** (preservation of central vision) may be found in some smaller occipital lesions (G in Fig. 2). Presumably, because of extensive macular representation in the occipital visual cortex, a smaller occipital lesion may spare macular function. A smaller lesion affecting only the inferior (E in Fig. 2) or superior (F in Fig. 2) optic radiations would create a contralateral **quadrantic homonymous deficit (or homonymous quadrantanopsia**) in the superior or inferior visual fields, respectively.

A **heteronymous hemianopsia is a visual deficit involving the nasal halves of both eyes, or the temporal halves of both eyes, which only occurs with a lesion of the optic chiasm** (B and C in Fig. 2). A lesion only affecting the optic chiasm at the midline would interrupt the decussating fibers from both nasal retinae, creating bilateral temporal visual field losses (B). Lesions compressing the outer optic chiasm on each side would affect only the temporal retinal fibers, creating bilateral nasal visual field deficits (C).

In summary, **optic nerve lesions produce scotomas or monocular blindness. Optic chiasm lesions produce heteronymous visual field defects. Lesions of the optic tract or optic radiations or occipital visual cortex produce homonymous visual field defects.**

2. Clinical syndromes of visual deficit

Acute unilateral optic nerve lesions are usually due to demyelination (multiple sclerosis in younger adults) or ischemia (in older adults). Multiple sclerosis may be initially manifest as **optic neuritis**. There typically is sudden blindness of part (scotoma) or all of one eye, which may feel achy or tender with eye movement. The pupils constrict poorly or not at all with light shined into the involved eye, but the pupils constrict normally with light shined into the normal eye. With an ophthalmoscope, the affected optic disc appears swollen with indistinct, blurry margins. (In some patients the inflammation is deeper or more posterior in the optic nerve, so the optic disc appears normal, despite visual impairment in that eye: retrobulbar neuritis.) Weeks to months later, vision has recovered completely or partially. In the latter case, a residual scotoma may be present, with or without impairment of visual acuity or color vision, possibly with a relative afferent pupillary defect (RAPD; see "Cranial Nerves, Brain Stem Reflexes, and Brain Stem Disorders"). The situation is now that of **optic atrophy**, and the optic disc appears more white or pale, with sharply defined edges. Although the examiner does not strictly observe atrophy here, some loss of ganglion cell axons has occurred. In general, the findings in one eye of visual deficit, abnormal pupillary light reflex and

swelling (or subsequent "atrophy") of the optic disc are due to lesions of the optic nerve or optic chiasm, and do not occur with visual pathway lesions posterior to the lateral geniculate body.

It is rare for acute optic neuritis to occur in both eyes simultaneously, so when both optic discs appear to be swollen and indistinct, this is usually due to **papilledema**. In papilledema, increased intracranial pressure affects both optic discs after a period of several hours, whether due to an extensive brain tumor or intracranial mass, meningoencephalitis, or pseudotumor cerebri (see "Headache"). Flame hemorrhages may be seen around the optic discs, and blood vessels passing through the optic disc may disappear from the plane of focus as they rise up over the "mound" of the elevated, swollen optic disc. Vision is not affected initially, but may be impaired or lost if the increased intracranial pressure persists without treatment. Patients often have headache, nausea and vomiting, or impaired consciousness from the elevated intracranial pressure.

A common lesion affecting the **optic chiasm** is a **pituitary tumor**, which arises from the sella turcica and exerts pressure on the center of the chiasm from below. Thus, the decussating, inferior, nasal retinal fibers would be affected first, creating a visual deficit in the superior temporal quadrants of the patient. A total lesion of the center of the optic chiasm would produce a bitemporal heteronymous hemianopsia. Of course, a pituitary tumor is likely to cause endocrine signs and symptoms as well.

Tumors or ischemic infarctions may cause lesions in the **optic radiations**. The inferior optic radiations bend anteriorly in the temporal lobe, and are also known as Meyer's loop. A lesion here causes a contralateral superior homonymous quadrantanopsia ("pie in the sky"). A lesion of the superior optic radiations in the parietal lobe causes a contralateral inferior homonymous quadrantanopsia ("pie on the floor"). **Cortical blindness** refers to the severe visual loss from bilateral occipital lobe lesions. This is usually a stroke syndrome from thrombosis of the distal basilar artery or emboli down its posterior cerebral artery branches. Although the patient is unable to see, the pupillary light reflex is intact and the optic discs appear normal since the retinal ganglion cells are not involved.

Auditory and Vestibular Systems

1. Bedside examination and related abnormalities

At the bedside or in clinic, hearing acuity is assessed crudely by whispering a number or word or holding a ticking watch near the patient's ear. Any detected hearing impairment may be due to conductive deafness or nerve (sensorineural) deafness. **Conductive deafness** is caused by impaired air conduction of sound stimuli, such as water or wax plugging up the external ear canal, or fusion or disruption of the bony ossicles. Simply put, conductive deafness is a system problem "before" the hair cell receptors. **Nerve (sensorineural) deafness** is caused by damage or impairment of the hair cell receptors or auditory nerve, as from drug toxicity or persistent exposure to loud noise. An **audiogram** provides precise identification of the type of deafness present, but requires special equipment in a sound-proofed room. Low-tone hearing loss is typical of conductive deafness, while high-tone hearing loss occurs with nerve (sensorineural) deafness. Severe or complete unilateral nerve deafness is usually due to an eighth cranial

nerve (CN VIII) lesion, since the more proximal, ascending auditory pathways are bilaterally represented.

Some bedside tests with a tuning fork (128 Hertz frequency "C") may also help determine the type of deafness present. Auditory stimuli are detected by bone conduction and air conduction. Bone conduction is the means by which vibrating sounds, such as one's own voice, are transmitted through the skull to sound receptors. Air conduction allows the detection of airborne sounds by the tympanic membrane and ossicles, which amplify the stimuli before sounds are transmitted to the cochlea. Because of this amplification system, **air conduction is normally more efficient than bone conduction**. The **Weber test** consists of holding the vibrating tuning fork at the top of the skull or middle of the forehead (Fig. 3). If nerve deafness has occurred in one ear, both air and bone conduction are impaired, so the vibrating tuning fork is heard better in the normal ear. If conductive deafness has occurred in one ear, the vibrating tuning fork is heard better in that deaf ear, since bone conduction is enhanced when room or environmental sounds are suppressed or reduced. (You can demonstrate this yourself. Plug one of your ears with your finger, and hold a vibrating tuning fork over your forehead. The plugged ear replicates conductive deafness, and you will hear the vibration better in it.). The **Rinne test** (Fig. 3) begins with holding a vibrating tuning fork at the mastoid bone, allowing the vibration to be heard by bone conduction. When no longer heard here, the vibrating tuning fork is then held outside the ipsilateral ear, where it should still be normally heard, since air conduction is better than bone conduction. If partial nerve deafness is present, air conduction is still better than bone conduction. In the case of conductive deafness, air conduction is no longer more efficient, so the vibrating tuning fork is no longer heard outside the ear once bone conduction at the mastoid bone ceases.

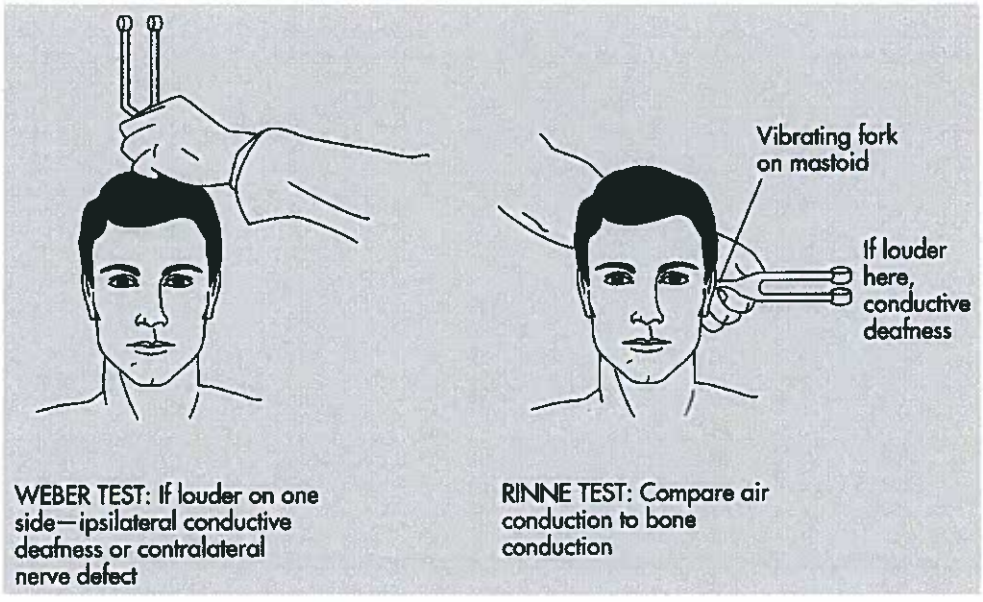

Fig. 3 Bedside hearing tests

Dysfunction of the vestibular system typically produces movement-induced dizziness or vertigo. An **electronystagmogram (ENG)** uses special equipment to record the eye movements and nystagmus induced by currents of warm or cool air entering the external ear canal, and determines whether the right or left vestibular system is impaired. This same oculovestibular reflex is also assessed by the cold caloric or ice water caloric test in comatose patients (see the Small Group Session "Coma and Brain Death"). Conscious patients, however, would not tolerate the noxious stimulus of ice water instilled in the external ear canal. A simple bedside test of the vestibular system is the **Dix-Hallpike maneuver** (Fig. 4). After explaining the procedure, the examiner helps the patient lie supine on the examination table, with his or her head tilted about 45 degrees below the edge of the table, turned to one side. This position selectively "tests" the posterior semicircular canal within the tilted, "lowered" ear. If this position creates rotatory nystagmus, that posterior semicircular canal is "overly sensitive" to head movement, and is likely the cause of positional vertigo. The patient is then helped to sit upright, and the head is tilted and lowered toward the other side for contralateral testing.

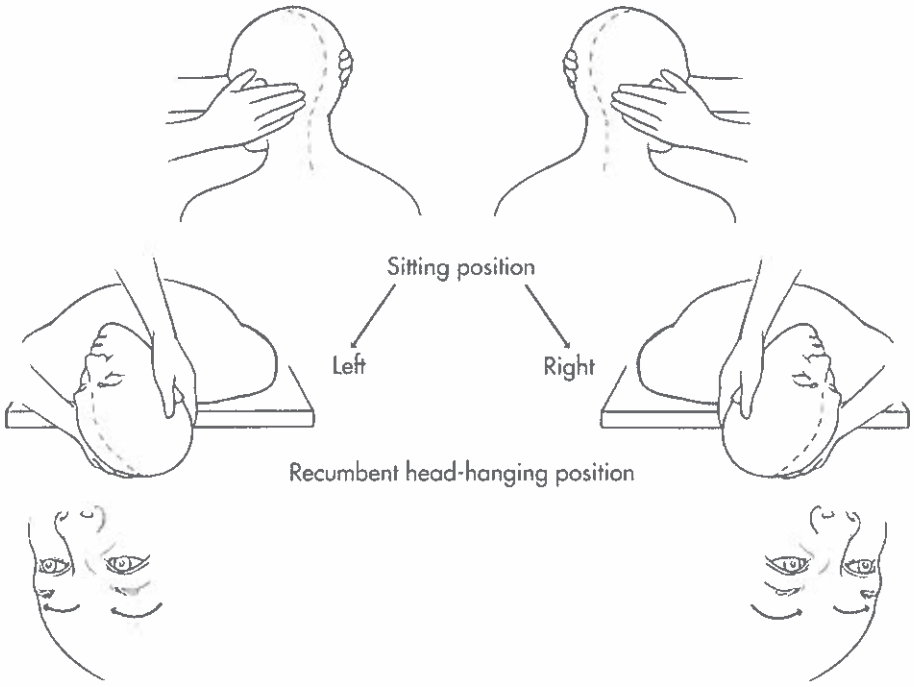

Fig. 4 Dix-Hallpike maneuver

Positional maneuver to test benign positional vertigo. The patient is moved rapidly from a sitting position to a recumbent, lateral, head-hanging position on the right or left. This induces vertigo and rotary nystagmus toward the affected ear when it is down. (From Collins RC: *Neurology*, Philadelphia, 1997, WB Saunders.)

2. Clinical syndromes of auditory and vestibular deficit

Causes of isolated hearing loss or deafness have already been mentioned with the discussion of conductive versus nerve deafness. Sudden, severe unilateral deafness may also be due to trauma, particularly with petrous bone fractures, or from ischemia in the territory of the anterior inferior cerebellar artery.

In **acute labyrinthitis**, the labyrinth of the inner ear may be affected by a viral infection or inflammation, causing severe vertigo with nausea and vomiting, hearing impairment, and unsteadiness of gait. Nystagmus, unilaterally decreased hearing, and gait ataxia are often found on examination. The symptoms resolve within days to weeks, and are temporarily helped with benzodiazepine, antihistamine, or antiemetic medication. Another clinically similar labyrinthine disorder is **Ménière's disease**, which consists of recurrent episodes of vertigo, deafness and tinnitus (abnormal buzzing or ringing in the ear). Here, the membranous labyrinth swells and ruptures, allowing potassium-rich endolymph to leak into the surrounding perilymph, disrupting the ionic gradient required for normal hair cell function. The clinical findings are similar to those of acute labyrinthitis, and the symptoms can be alleviated by the same medications already mentioned. However, repeated episodes of Ménière's disease may lead to complete, permanent deafness. Dietary salt restriction and diuretic medication may help reduce production of endolymph and lessen the risk of chronic deafness.

Benign positional vertigo is a common problem of elderly patients. With aging, degeneration of otoliths and displaced calcium crystals and other debris can lodge around the cilia of semicircular canal hair cells, making them "oversensitive" to minor movements of the head. Patients complain of transient but annoying vertigo when standing up or turning their heads. The Dix-Hallpike maneuver is a helpful diagnostic test, and similar head-turning exercises may be used therapeutically to either disperse the troublesome debris in the semicircular canals or help the patient readapt to head movements.