CHAPTER 23
Visual System

INTRODUCTION

Of all our senses, vision is the most important: we perceive the world mostly through our eyes. Even though light intensity varies by a factor of 10 million between the brightest snowy day and a starlit night, our eyes and visual system adapt to these intensity changes. We can discriminate between thousands of hues and shades of color. Our eyes are set in our heads in such a way that each eye sees almost the same visual field, making depth perception possible.

In the visual system the primary, secondary, and tertiary neurons are in the retinae and are all part of the central nervous system. The right field of vision projects to the left cerebral hemisphere; the left field of vision projects to the right cerebral hemisphere.

STRUCTURE OF THE EYE

The anatomy of the receptor organ, the eye is shown in *Figure 23-1.* It has three layers, or tunics:

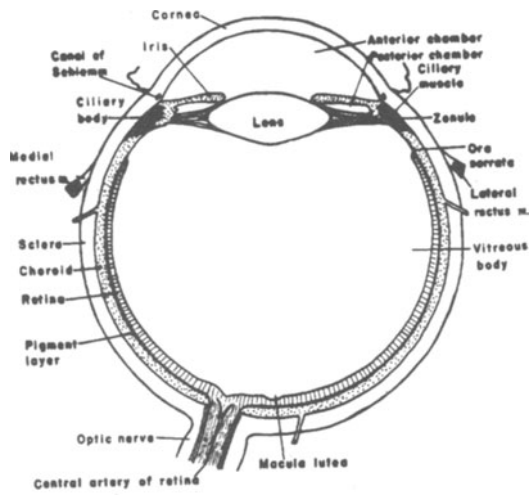


Figure 23-1A. Horizontal meridional section of the eye. (From Leeson and Leeson. 1970. Histology, Philadelphia, W.B.Saunders.)

1. The outer fibrous tunic - cornea and sclera.

2. The middle vascular and pigmented tunic - choroid, ciliary body, iris and pupil.

3. The inner neural tunic - retina with pigmented epithelium and neuronal layers.

The **outer fibrous tunic** consists of two parts: the anterior transparent cornea and the posterior fibrous white sclera.

The **cornea,** the window to the world, allows light rays to enter the eye. Most of the refraction needed to focus light rays on the retina occurs at the air-cornea junction. The sclera forms the white of the eye and the rest of the outer covering.

Middle Tunic- the choroid, the ciliary body, and the iris. The middle tunic is richly vascular and provides oxygen and nutrients to the inner, photoreceptor layer, and the retina.

The choroid is vascular and pigmented and forms the posterior portion of this tunic. Its inner portion is attached to the pigmented layer of the retina. The ciliary body is found in the anterior portion of the middle tunic and consists of a vascular tunic and the ciliary muscle. The ciliary body surrounds the lens and consists of a vascular tunic (the ciliary muscle) and the suspensory ligaments (the zonule), which suspend the lens *(Fig 23-1B)*. The ciliary muscles consist of meridional and circular fibers. The meridional fibers are external to the circular fibers. The ciliary muscles are the muscles used in accommodation, focusing on near objects.

Lens. The lens separates the anterior chamber from the vitreous body and completes the refraction of the entering light. A fibrous network, the zonule, suspends the lens. For distance vision the fibers are taut and the anterior surface of the lens is pulled flat. For close vision the ciliary muscle contracts,

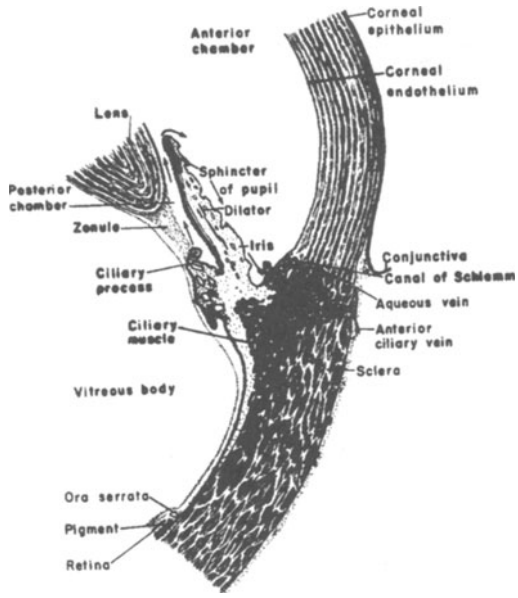


Figure 23-1B.Enlargement of a potion of the meridional section to show the angle of the eye. The letter P indicates the pectinate ligament or the trabecular meshwork. The arrows indicate the course of circulation of aqueous fluid. (From Leeson and Leeson, Histology, Philadelphia, WB Saunders, 1970.)

the fibers go slack, and the anterior surface becomes more convex; the eye accommodates. A decreased ability to focus on close objects is a normal consequence of aging. The term "visual acuity" refers to the resolving power of the eye in terms of the ability to focus on near or distant objects. For example, 23/40 means the patient's eye sees at 23 feet what the normal eye sees at 40 feet.

The pigmented iris on the anterior portion of the vascular tunic divides the space between the lens and the cornea into an anterior chamber (between the lens and the cornea) and a narrow posterior chamber (between the suspensory ligaments of the lens and the iris). The iris is a circular structure that acts as a diaphragm to control the amount of light falling on the retina. The opening is analogous to the f-stop of a camera. The eye has a range equivalent to the range from f2 to f22.

PUPILLARY REFLEXES

Pupillary Muscles. Two sets of muscles in the iris control the size of the pupil, sphincter and the dilator pupillae. When the circular muscles, the sphincter pupillae contract, the iris is drawn together and the pupil constricts, such as purse strings close the mouth of the purse. The second set of muscles, the dilator pupillae, are radial and draw the iris back toward the sclera.

The sphincter pupillae are supplied by the parasympathetic nervous system via the ciliary nerves, the fibers of which run together with nerve III. Transmission by this pathway is cholinergic, the dilator pupilla are supplied by the sympathetic nervous system via the superior cervical ganglion. Neurotransmission in this pathway is alpha-adrenergic.

Pupillary Reflexes. When light is flashed into either eye, the pupil dilates, the light reflex. When the eye focuses on close objects, accommodation occurs

1. *Light Reflex.* In dim light the aperture of the pupil increases (dilator muscle) while in bright light the aperture of the pupil decreases (sphincter muscle). The afferent fibers arise in the retina and travel with the optic nerve and tract, these fibers pass through the lateral geniculate and synapse in the pretectal region of the midbrain. Neurons of the pretectal nuclei project bilaterally to the Edinger-Westphal nucleus.
2. *Accommodation.* This reflex changes the refractive power of the lens by the ciliary muscle contracting and decreasing the force on the suspensory ligament of the lens; the lens assumes a more rounded appearance with a shorter focal length. The neural pathway arises in the occipital lobe and the final efferent fibers run together with nerve III. In looking at distant objects the pupil dilates while in focusing on nearby objects the pupil constricts.
3. *Fixed Pupil.* In the absence of trauma to the eye, a dilated pupil that does not respond to light, fixed, is usually a sign of pressure on the third nerve. This is typically from supratentorial mass lesions that have produced herniation of the mesial segments of the temporal lobe through the tentorium. Compression of the third nerve and of the

midbrain occurs. Progressive rostral- caudal damage to the brain stem then evolves. This problem when first detected must be treated as a neurosurgical emergency.

Inner Tunic, Retina - pars optica, pars ciliaris, and pars iridica. The inner tunic contains photoreceptor cells and nerve cells. They are organized so that photoreceptor cells containing the photopigments are closet to the sclera and the nerve cells are above them. The nerve cells send fibers to the optic nerve through a pigment-free area, the optic disc. Light must traverse the nerve network to reach the photoreceptors.

The pars ciliaris and iridica are primarily a pigmented region on respectively the ciliary body and iris. The pars optica contains the photoreceptor cells and will be the focus of our discussion.

The pars optica contains:

- *outer pigmented layer,*
- *inner photoreceptor cells closest to the sclera,*
- nerve cells that are internal to photoreceptors and form the optic nerve that leaves the retinae through a pigment-free area, the optic disk. Light must traverse the nerve network to reach the photoreceptors.

The pars optica of the retina consists of the following layers *(Fig.23-2A):*

1. Pigmented epithelium (most external),
2. Receptor cell layer - rods and cones,
3. External limiting membrane,
4. Outer nuclear layer containing the nuclei of rods and cones,
5. Outer plexiform layer- containing the synapses of rods and cones with the dendrites of bipolar and horizontal cells, and the cell bodies of horizontal cells,
6. Inner nuclear layer- containing the cell bodies of bipolar, and amacrine cells,
7. Inner plexiform layer- containing the synapses of bipolar and amacrine cells with ganglion cells,
8. Ganglion cell layer,
9. Optic nerve layer, and
10. Internal limiting membrane.

Photoreceptors - Rods and Cones.

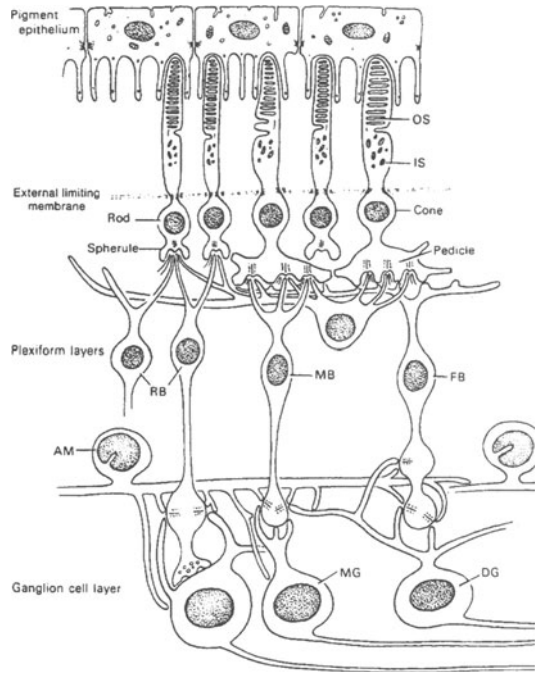
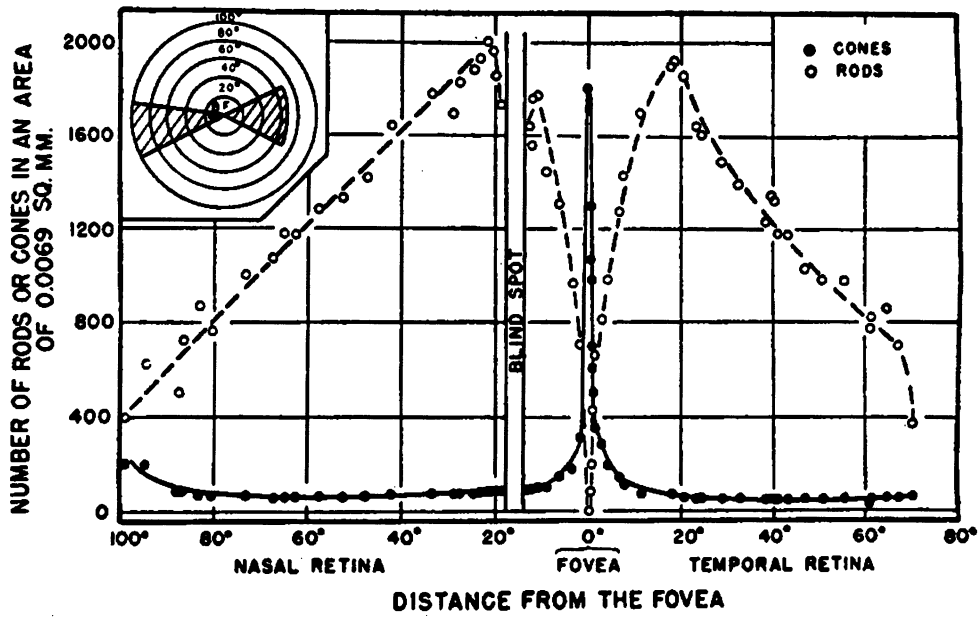


Figure 23-2 A. Schematic diagram of the ultrastructural organization of the retina. Rods and cones are composed of outer (OS) and inner segments (IS), cell bodies, and synaptic bases. Photopigments are present in laminated discs in the outer segments. The synaptic base of a rod is called a spherule; the synaptic base of a cone is called a pedicle. Abbreviations: RB = bipolar cells, MB=midget bipolar cell, FB a flat bipolar cell, AM = amacrine cell, MG = midget ganglion cell, DG = diffuse ganglion cell. (From Carpenter, M.B.A Core Text of Neuroanatomy Baltimore, Williams and Wilkins).

There are two types of photoreceptor cells: rods and cones.

The *rods* are sensitive in dim light as they contain more of the photosensitive pigment rhodopsin than the cones. This system has poor resolution; newspaper headlines are the smallest letters that can be recognized.

The *cones* are sensitive to color. There are there separate groups of cones each of which contains photopigments that are primarily sensitive 1) to blue - short wave lengths, 2) to green - middle wave lengths, or 3) to red longer wave lengths. Color vision requires light levels greater than bright moonlight and has high resolution-fine detail can be seen.
The rods and cones are not uniformly distrib-



uted in the retina. Most of the 6 million cone cells are located in an area 2 mm in diameter, the macula lutea (Fig. 23-1,), which can be seen through the ophthalmoscope. In the center of the macula lutea lies a zone of pure cones, the fovea *(Fig. 23-2B)*. The rest of the macula lutea is composed of both rods and cones, and most of the peripheral retina contains only rods. There are about 123 million rods.

Figure 23-2B. The densities of cones and rods on or near the horizontal meridian through a human retina. The insert is a schematic map of the retina showing the fovea (F) and the blind spot (B). The striped area represents the regions of the retina, which were sampled in obtaining the counts plotted here. From Chapanis after Osterberg, Acta Opth, 1935, Suppl.6 (Blackwell).

Rods - Vision in Dim Light and Night Vision

The rods permit vision, *scotopic* (dark vision), in the dark-adapted eye. Rods contain a single pigment, *rhodopsin*, which is related to vitamin A_1 (retinol). The spectral sensitivity of night or scotopic vision is identical to the absorption spectra of rhodopsin (Fig 23-3) Light that can be absorbed by rhodopsin is seen; light that cannot be absorbed such as red light is not seen at these low light intensities. Scotopic vision has quite poor definition; two light sources must be quite far apart to be distinguished as two sources rather than one. Peripheral vision has progressively poor definition. The modern human uses this system very little since we have bright, portable light sources or radar. Several modern inventions,

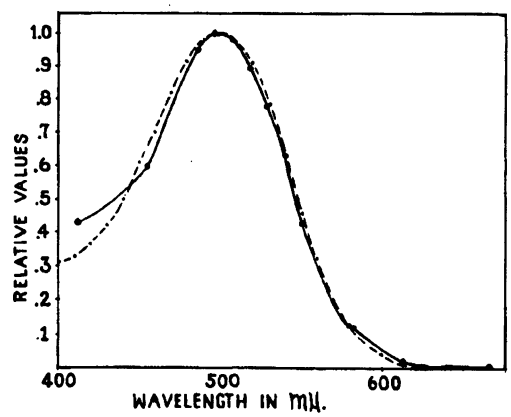


Figure 23-3. Comparison of the absorption curve of pure visual purple (interrupted line) with the intensity of light just visible in a dark-adapted human subject (continuous curve). From Ludvigh, Arch.Ophthal.20: 713.1938 (AMA).

such as night goggles with infrared sensors, are designed to compensate for these limitations.

The visual pigment, rhodopsin absorbs photons. Each rhodopsin molecule consists of

retinal and opsin. Retinal absorbs the light while opsin is the protein in the plasma membrane of the rods. The photon converts retinal from the 11-cis form to the all-trans form. The all-trans retinal has to be re isomerized to 11-cis retinal to reform rhodopsin and to begin the process again. The photons result in hyperpolarization of the plasma membrane that is the light dependent part of visual excitation. The photoreceptors are depolarized in the dark due to the sodium channels remaining open, called the dark currents. The dark current is turned off in light. Daylight prevents the regeneration of rhodopsin in the rods.

After 5-10 minutes in the dark, scotopic vision begins to return as the rhodopsin is regenerated, and reaches maximal sensitivity after 15 to 23 minutes. Since rhodopsin does not absorb red light, using red goggles can preserve night vision.

Cones - Color Vision

Color vision requires much higher light intensities and occurs primarily when the image is focused upon the macula. Each cone contains one of the three-color pigments whose absorption spectra are shown in *Figure 23-4.* The visual pigment of the cones consists of opsin and retinal.

The cones are divided into three separate groups that contain photopigments primarily sensitive to a different part of the visible spectrum as follows:

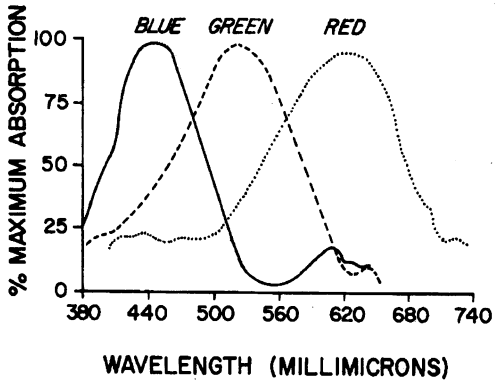


Figure 23-4. The absorption spectra of three types of cones. (Drawn from records in Marks, Dobelle, and MacNichol. 1964. Science 143:1181; and Brown and Wald. 1964. Science 144:45.)

1. Blue -423 nm (short wavelengths),
2. Green -530 nm (middle wave lengths), and
3. Red - 560 nm (longer wavelengths).

Color vision requires light levels greater than bright moonlight and has high resolution so that fine detail can be seen. Any color that does not excite two pigments, such as deep purple (400 nm) and deep red (650 nm) will be hard to discriminate. For this reason, a color of 660 nm in bright light can mimic a color of 640 nm in weaker light. To distinguish the color orange, 600 nm, the visual system compares the relative absorption by two visual pigments, in this case red and green.

Comparing the absorption by one visual pigment to the overall brightness apparently makes these color discriminations. For this reason a color of 660nm, in bright light, mimics a color of 640 nm in weaker light. Only when the two types of cones are excited can color and brightness be determined independently.

Electrophysiology of Retinal Photoreceptors and Neurons.

Synaptic Organization of the Rods, Cones and Neurons in the Retinae (Figure 23-2A)

1. Rods and cones synapse on the dendrites of bipolar cells and horizontal cells. In the peripheral portion of the retina, 60 degrees from the center, as many as 600 rods may converge on single bipolar cell, while in the fovea, only one or two cones may do so. In the macula lutea both rods and cones may innervate a single bipolar cell.

2. Axons of bipolar cells and amacrine cells synapse on dendrites of ganglion cells. Once again, the degree of convergence on the ganglion cell depends on the region of the retina. Ganglion cells in the fovea connect with only one bipolar cell.

3. Axons of ganglion cells leave the retina; form the II cranial nerve that synapses in the lateral geniculate, hypothalamus, and midbrain. There are about a million optic nerve fibers. Clearly, considerable data reduction has occurred between the 100 million rods and

cones and the one million optic nerve fibers.

Two other types of neurons are found in the retina: horizontal and amacrine cells. Horizontal cells are found in the outer plexiform layer (next to the rods and cones); their dendrites make contact with numerous rods and cones over a 230 to 400-μM field. Their axons run transversely along the retina and branch to make contact with many other rods or cones at the bipolar-cell junction. The horizontal cells connect groups of visual receptors in one area with groups in another area. Amacrine cells lie in the inner plexiform layer (next to the ganglion cells). The dendrites form postsynaptic contacts with bipolar cells and presynaptic contacts with ganglion cells; no axon has been described.

Response of the cells to light *(Fig. 23-5)*

Rods and Cones. Both rods and cones show a hyperpolarizing response to light. This very surprising result has been obtained in all vertebrate visual receptors studied to date. Contrary to what one would expect, they act as if dark were the stimulus. During darkness, high concentrations of cyclic GMP increase membrane conductance and give rise to a dark current that flows from the outer to inner segment, depolarizing the cell. After absorbing a photon, rhodopsin activates cyclic-GMP-phosphodiesterase, which cleaves cyclic GMP, lowers cyclic GMP levels, and by reducing the membrane conductance, hyperpolarizes the rod. Also contrary to expectation, the rod and cone cells release transmitter when it is dark, but this release ceases when it is light.

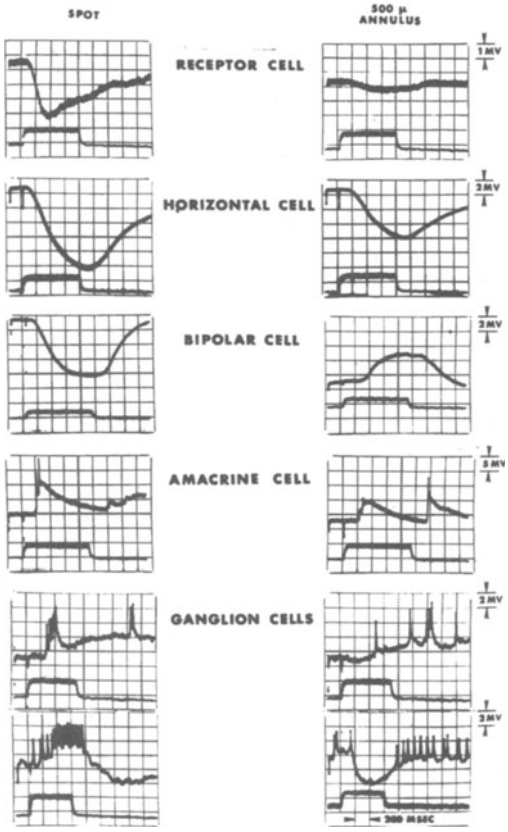


Figure 23-5. Responses of the cell types in the Necturus retina. From F.S. Werblin and J.Dowling. 1969. J. Neurophysiol. 32:339 (Amer. Physiol. Soc.)

Horizontal Cell of retinae - hyperpolarizes in response to light, but it has a large receptive field. Even when the light is shining on receptor cells 250μm away, there is a strong hyperpolarizing response. Potential spread through the horizontal layer is by slow potentials; no action potentials have been observed.

Bipolar Cell: Both the direct hyperpolarizing receptor response and the horizontal-cell output impinge on the bipolar cell. Here we see the first type of data reduction carried out in the retina: center surround contrast enhancement. The bipolar cell has one type of output--hyperpolarization--for a bright spot on a dark field and another type of output--depolarization--for a large bright field.

Amacrine Cell. Recordings from the amacrine cell show that it responds primarily to sudden changes in light intensity. Note that it is active primarily when the light turns on or off. This is the second type of processing, or data reduction, that takes place in the retina: dynamic detection.

Ganglion Cells. There are two types of ganglion-cell responses –amacrine or bipolar cell.

Amacrine Cell Response. This response is, primarily a dynamic response to turning the light on or off. Some cells react more strongly to the light coming on than off; others have the opposite response. These cells are inhibited by illumination of both a spot and the surrounding area.

Bipolar Cell Response. The second type of ganglion-cell response reflects bipolar-cell activity. When only the central portion of its receptive fields is illuminated, the ganglion cell responds with an intense train of action potentials during the illumination that abruptly stops when the light ceases. The frequency of the action potentials during illumination is proportional to the logarithm of the brightness of the light. When both the spot and the surrounding area are illuminated, there is less activity than occurs in the dark (the background level). When only the surround is illuminated, there is no specific response.

A third type of ganglion cell response (not shown in figure 23 –5)- is a center off type. When the center of the receptive field is illuminated, action potentials frequency decreases. When the surround is illuminated activity is greater than the background level, but not as intense as in the second type of response to a spot noted above.

To summarize ganglion cells signal the onset and cessation of light flashes and are effectively stimulated only by edges or abrupt transitions between light and darkness in their fields. Diffuse illumination does not stimulate ganglion cells very well.

The axons from the ganglion cells traverse the inner surface of the retina to the optic disk, where they turn and form the optic nerve, which runs for about 1 inch before entering the optic foramen on its way toward the diencephalon.

Optic Nerve. Electrical recordings from optic nerve fibers also show the types of data reduction occurring in the retina. First there is convergence; many receptors must fire one bipolar cell in the fovea Consequently the "grain"of the visual image depends on the location on the retinae. Second brightness can be detected. Third there is a center surround contrast enhancement. Lastly there is a dynamic or motion detection.

A spot of light on the retinae will be coded in terms of its brightness and its edges. As it is turned on and off, the dynamic receptors fire to alert higher centers. The spot of light will receive much more attention in the optic nerve bandwidth if it is moved around because each tiny movement will set of a new set of dynamic or movement receptors and these receptors make up about half the total optic nerve fibers. For example the rotating flashing lights of emergency vehicles attracts more attention than a fixed light.

VISUAL PATHWAY *(Fig.23-6)*

1. Origin of Cranial Nerve II from gan-

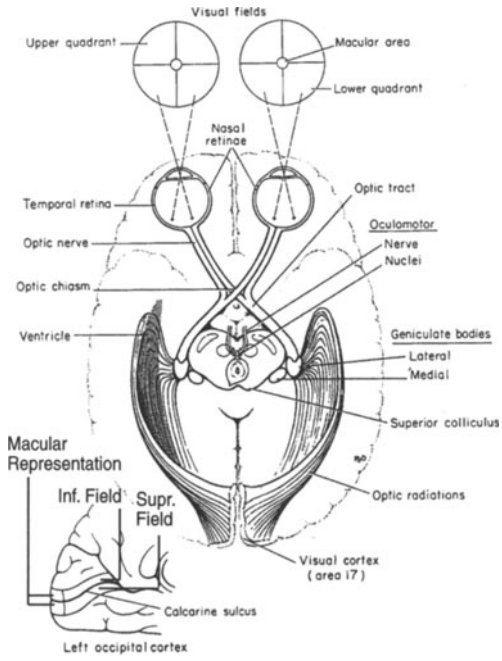


Figure 23-6. The visual pathways viewed from the ventral surface of the brain. Light from the upper half of the visual field falls on the inferior half of the retina. Light from the temporal half of the visual field falls on the nasal half of the retina; light from the nasal half of the visual field falls on the temporal half of the retina. The visual pathways from the retina to the striate cortex are shown. The plane of the visual fields has been rotated 90 degrees toward the reader. The inset (lower left) shows the projection of the quadrants of the right visual field on the left calcarine (striate) cortex. The macular area of the retina is represented nearest the occipital pole. Fibers mediating the pupillary light reflex leave the optic tract and project to the pretectal region; fibers from the pretectal olivary nucleus relay signals bilaterally to the visceral nuclei of the oculomotor complex. (Modified from Carpenter and Sutin. 1983. Human Neuroanatomy. Courtesy of Williams and Wilkins.)

glion cell layer in the retina,

2. Optic nerve,

3. Optic chiasm,

4. Optic tract,

5. Synapse in lateral geniculate nucleus of the thalamus,

6. Visual radiation from lateral geniculate nucleus to the striate (visual) cortex - area 17 of the occipital lobe,

7. Visual association cortex - areas 18 and 19, and

8. Multimodal associations with cortical regions in the posterior temporal-parietal, and frontal lobes in both hemispheres (Fig. 18-11).

VISUAL FIELDS

Retina.

Each retina is divided into a temporal or nasal half by a vertical line passing through the fovea centralis. This also serves to divide the retina of each eye into a left and right half. A horizontal line passing through the fovea would divide each half of the retina and macula into an upper and lower quadrant. Each area of the retina corresponds to a particular sector of the visual fields, which are named as viewed by the patient. For each eye tested alone, there is a left and right visual field corresponding to a nasal and temporal field. Because of the effect of the lens, the patient's left visual field projects onto the nasal retina of the left eye and the temporal retina of the right eye. Similarly, the upper visual quadrants project to the inferior retinal quadrants.

Optic Nerve and Optic Chiasm.

At the chiasm, the fibers in the optic nerve from the left nasal retina cross the midline and join with the fibers from the right temporal retina to form the right optic tract. This dense bundle of over a million fibers runs across the base of the cerebrum to the lateral geniculate nucleus of the thalamus. Fibers from the inferior nasal retina (the superior, temporal visual field) cross in the inferior portion of the chiasm, which usually lies just above the dorsum sellae.

By this crossing, all the information from the left visual field is brought to the right lateral geniculate nucleus and subsequently to the right calcarine cortex. The fibers of the right nasal retina also cross in the chiasm to join the fibers from the left temporal retina. Thus, the visual pathway repeats the pattern in other sensory systems; the right side of the visual field is represented on the left cerebrum.

Lesions in the Optic Pathway:

1. In the retina or the optic nerve before the optic chiasm (2 in *Figure 23-7*) there is no sight in that eye - monocular blindness.

2.In the chiasm, the fibers from both temporal visual fields are cut; the result is bitemporal hemianopsia (3 in Figure 23-7)

3. Lesions behind the optic chiasm produce blindness in one visual field, a homonymous hemianopsia from injury to the optic tract, the lateral geniculate body or the genicu-

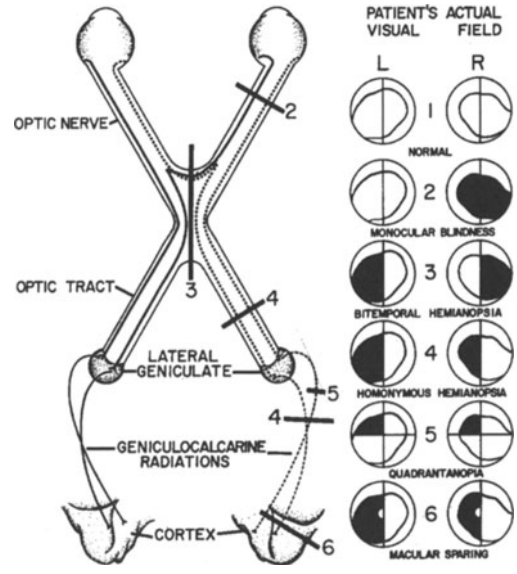


Figure 23-7. Common lesions of the optic tract.1) Normal, 2) optic nerve, 3) optic chiasm, 4) optic tract or complete geniculate or complete geniculocalcarine radiation or complete cortical 5) temporal segment of geniculocalcarine radiation (Meyer's loop) producing a superior quadrantanopia. If the parietal segment of this radiation were involved, an inferior quadrantanopia would result. 6) Calcarine or posterior cerebral artery occlusion producing a homonymous hemianopia with macular sparing.

localcarine radiation (4 in Figure 23-7).

4. Partial lesions of the geniculocalcarine radiations produce a quadrantanopsia (5 in figure 23-7).

5. Macular sparing is seen with lesions in the visual cortex following calcarine artery infarcts (6 in Figure 23-7). The macula is represented close to the occipital pole and this area receives anastomotic flow from the middle cerebral artery.

The following case is an example of a lesion in the optic nerve, anterior to the optic chiasm.

Case History 23-1: This 53-year-old white right-handed housewife had a progressive 23-year history of right-sided supraorbital headache with decreasing acuity and now almost total blindness in the right eye. During the 3 years before admission, intermittent tingling paresthesia had been noted in the left face, arm, or leg. About 1 year before admission, the patient had a sudden loss of consciousness and was amnestic for the events of the next 48 hours. No explanation for the episode was clearly established. The cerebrospinal fluid protein was reported to be elevated (230 mg/dl). The patient and her family reported some personality changes over a period of several years, including a loss of spontaneity and increasing apathy.

General Physical Examination: There was a minor degree of proptosis (downward protrusion of the right eye).

Neurologic Examination: *Mental Status:* The patient was, in general, alert but at times would become lethargic. Her affect was flat. At times, she would laugh or joke in an inappropriate manner. *Cranial Nerves:* There was anosmia for odors, such as cloves, on the right and a reduced sensitivity on the left. Marked papilledema (increased intracranial pressure with elevation of the optic disk and venous engorgement was present in the left eye. In contrast, there was pallor of the right optic disk indicating optic atrophy. Visual acuity in the right eye was markedly reduced. This combination of funduscopic findings is termed the Foster Kennedy syndrome. The patient had only a small crescent of vision in the temporal field of the right eye; only vague outlines of objects could be seen. A slight left central facial weakness was present. *Motor System:* movements on the left side were slow. *Reflexes:* A release of grasp reflex was present on the left side.

Clinical diagnosis: Subfrontal meningioma rising from the inner third sphenoid wing. Alternative location would be olfactory groove.

Laboratory data: *Imaging studies* demonstrated a tumor blush in the right subfrontal region extending back to the right optic nerve groove consistent with a meningioma arising from the olfactory groove or inner third sphenoid wing, *(Fig. 23-8)*

Hospital Course: A bifrontal craniotomy performed by DR. Sam Brendler exposed a well-encapsulated smooth tumor a meningioma attached to the inner third of the sphenoid wing. Approximately 90 to 95% of the tumor was removed exposing the right optic nerve. Examination 4 months after surgery indicated right anosmia and right optic atro-

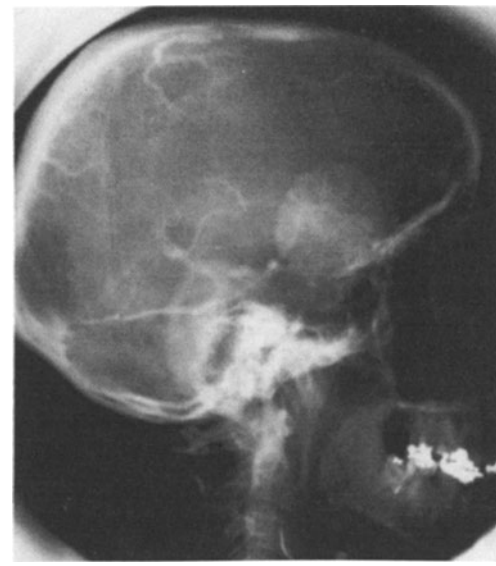


Figure 23-8. Sphenoid-wing meningioma producing compression of the right optic and olfactory nerves. Case 23-1 Refer to text. Right carotid arteriogram, venous phase demonstrating tumor blush in the subfrontal area of the anterior fossa, extending into the middle fossa. (Courtesy of Dr. Samuel Wolpert. New England Medical Center Hospitals.)

phy were present.

Optic Chiasm: A bitemporal defect results *(Fig. 23-9, 23-10, 23-11)*. This may be a bitemporal hemianopsia or an incomplete bitemporal field defect. The usual cause is a pituitary adenoma or a supra sellar tumor such as a craniopharyngioma.

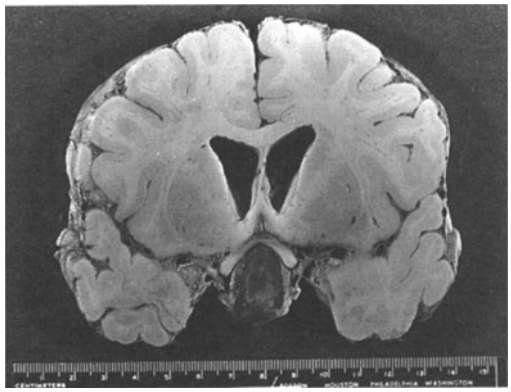


Figure 23-9. A large pituitary adenoma with secondary hemorrhage has extended upward out of the sella to compress the optic chiasm and optic nerves. This 53-year-old white male 11 months before death experienced difficulty reading small print, with greater involvement of the right eye than the left. This progressed to total blindness in the right eye and shortly thereafter-sudden loss of vision in the left eye, an absence of pupillary responses, and a bilateral anosmia. Vision improved after partial surgical removal and irradiation, but fatal meningitis developed. (Courtesy of Drs. John Hills and Jose Segarra).

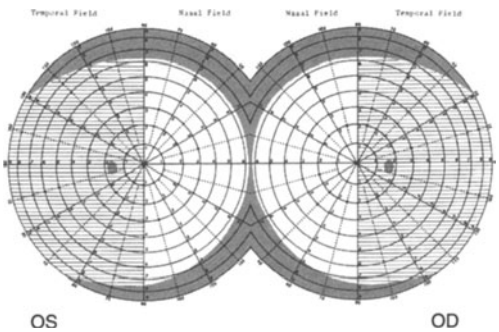


Figure 23-10. Pituitary adenoma. Visual fields demonstrating a bitemporal hemianopia. O.S. = left eye; O.D. = right eye. This 51-year-old obese male with large puffy hands and a prominent jaw, declining sexual interest for 18 years, an 8-year history of progressive loss of visual acuity, a 6- to 7-month history of diplopia due to a bilateral medial rectus palsy and headache had stopped driving because he was unable to see the sides of the road. Urinary adrenal and gonadal steroids and thyroid functions were low, with no follicle-stimulating hormone. Imaging studies demonstrated a large pituitary adenoma with significant suprasellar extent.

Case 23-2. Patient of Dr. Martha Fehr. This 45-year-old man had an 18-month history of a progressive alteration in vision. He was unable to see objects in the right or left periphery of vision. He was concerned that in the process of driving he might hit pedestrians stepping off the sidewalks in the periphery of vision. He had also experienced over 6-8 months, progressive headaches, loss of energy and libido.

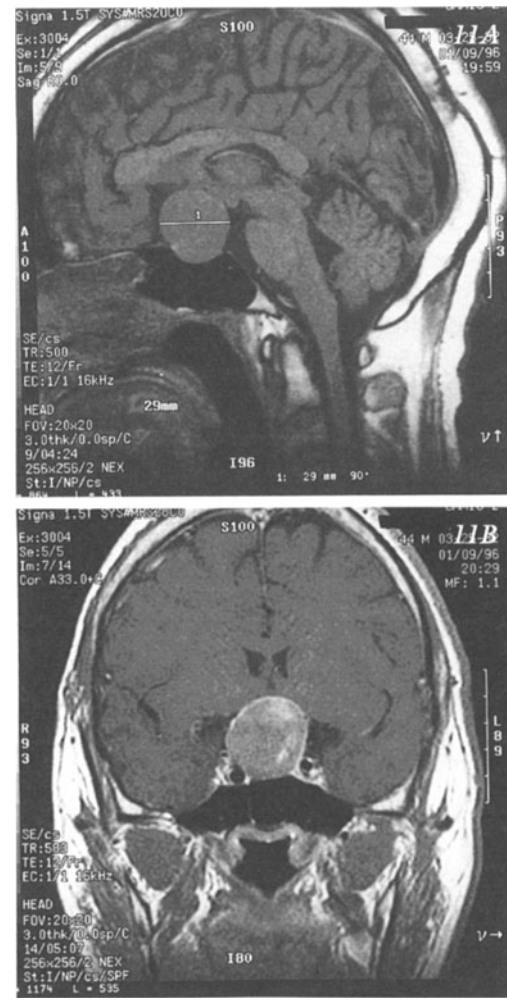


Figure 23-11. A large pituitary adenoma in a 44-year-old man with a bitemporal hemianopia. Case 23-2. Significant extrasellar extension compresses the T1 optic chiasm. MRI scans: A) midline sagittal section B) coronal section. (T1 with contrast.)

Neurologic exam: Normal except for a bitemporal hemianopia.

Clinical diagnosis: Pituitary adenoma compressing the optic chiasm.

Laboratory data: *Endocrine studies:* all were normal. *MRI:* A large macroadenoma measuring 3 cm in diameter extended outside of the sella turcica to compress the optic chiasm (23-11).

Subsequent course: Dr. Gerald McGuillicuddy performed a gross total trans sphenoidal resection of the tumor, with a significant improvement in vision. Three years later, the patient again had visual symptoms and eye pain. MRI scans indicated regrowth of tumor with possible impression on the right optic nerve. He was treated with radiotherapy.

Note that males are more likely to present with large pituitary adenoma compressing the optic chiasm. Women are more likely to present with microadenomas or small macroadenomas since they are more likely to be seen at an earlier stage due to an initial complaint of amenorrhea.

Lateral Geniculate Nucleus (LGN).

The LGN is primarily a relay station in the visual pathway. Some optic fibers for pupillary light reflexes bypass the LGN and descend into the pretectal region of the midbrain. Others, for coordinating eye and head or neck movements, descend to the superior colliculus. Also some of the retinal fibers synapse in the suprachiasmatic nucleus of the hypothalamus and then project to the pineal by way of the sympathetic fibers where they influence circadian rhythms and endocrine functions.

The lateral geniculate nucleus is a six-layer horseshoe-shaped structure. Visual processing begins in the large and small cells in the LGN. The small cells are in layers 1 to 4, and the large cells in layers 5 and 6. Each LGN cell receives direct input from a specific area of the retina, and each area of the LGN is driven from a specific area of the visual field. Each receptive field has a center that is either on or off and a surround that has the opposite polarity.

The optic nerve fibers from the temporal visual field, the crossed fibers, end in layers 1, 4, and 6. The fibers from the temporal retina (the nasal visual field), the uncrossed fibers, end in layers 2, 3, and 5. Each optic nerve fiber ends in one layer on 5 or 6 cells. The total number of cells and fibers is roughly equal. LGN cells respond well to small spots of light and to short, narrow bars of light, as well as to on-off stimuli. Diffuse light is a very poor stimulus.

Cells in the LGN grow rapidly in the first 6 to 12 months after birth, under the influence of visual stimuli. Visual sensory deprivation during this period leads to atrophy and lifelong blindness. Two major streams of visual information leave the retina to the LGN (1) The magnocellular stream starts in the large cells of the retinal ganglion (Y-cells) that project to the magnocellular layers (layers 1 and 2) which subsequently project to the striate cortex. The cortical neurons of the magnocellular stream respond to movements, orientation and contrast but respond poorly to color. (2) The parvocellular stream begins with small cells of the retinal ganglion layer (X-cells) that project to parvocellular layer of LGN layers 3 and 6. These cells respond either to shape and orientation or color red/green and blue/yellow.

Optic Radiation.

The fibers to the visual cortex leave the lateral geniculate nucleus and form the optic radiation, which sweeps around the posterior horn of the lateral ventricle, through the parietal lobe to the occipital lobe and the calcarine cortex (Fig. 23-6). Some fibers from the lateral geniculate nucleus also enter the inferior pulvinar nucleus.

The visual radiation is so extensive that not all the fibers can pass directly posterior to the calcarine cortex. Only the most dorsally placed fibers pass back deep into the parietal lobe. The more ventrally placed fibers pass forward (Myer's loop) and deep into the temporal lobe, swing posteriorly around the anterior portion of the inferior horn of the lateral ven-

tricle, and continue posteriorly lateral to the ventricle wall to the calcarine cortex. In contrast to the densely packed optic tract, the optic radiation fans out widely on its passage through the parietal and temporal lobes.

OCCIPITAL LOBE

The occipital lobe in Brodmann's numerical scheme consists of areas 17, 18, and 19. From a cytoarchitectural standpoint, area 17 represents a classic example of specialized granular cortex, or koniocortex. Areas 18 and 19 represent progressive modifications from koniocortex toward homotypical cortex (Refer to chapter 17 for a discussion of cytoarchitecture).

Area 17, the striate cortex, the principal visual projection area in humans is found primarily on the medial surface of the hemisphere, occupying those portions of the cuneus (above) and lingual gyrus (below) that border the calcarine sulcus. For this reason, it is often termed the calcarine cortex. Much of this cortex is located on the walls and in the depths of this sulcus.

The extrastriate visual cortex, including parastriate areas 18 and 19 forms concentric bands about area 17 and is found on both the medial and lateral surfaces. Area 19 in humans extends onto the adjacent mid and inferior temporal gyri.

Most of our understanding of occipital-lobe function comes from simian studies. In non-human primates five visual areas have been identified:

V1 - corresponding to area 17 in humans,
V2 and V3 - corresponding to area 18, and
V4 and V5 - corresponding to area 19.

Area 17 is the primary visual area, and receives the termination of the geniculocalcarine (or optic) radiation. This projection is arranged in a topographic manner, with the superior quadrant of the contralateral visual field represented on the inferior bank of the calcarine fissure, and the inferior quadrant of the contralateral visual field represented on the superior bank. The macula has a large area of representation, which occupies the posterior third of the calcarine cortex and extends onto the occipital pole.

Area 17 receives fibers from and sends fibers to area 18 but does not have any direct callosal or long-association-fiber connections to other cortical areas. The columnar and horizontal organization of the primary visual cortex has been discussed previously in chapter 17. Area 17 is more mature at birth and has the most precise map. The other visual areas develop through maturation and experience, although experience modifies V1 as well (see Chapter 29).

Area 18 has extensive connections with areas 18 and 19 of the ipsilateral and contralateral hemispheres, which were demonstrated by both the earlier strychnine neuronography studies and the more recent horseradish peroxidase studies.

Callosal fibers enter the opposite hemisphere, and association fibers communicate with the premotor and inferior temporal areas and area 7 of the adjacent parietal cortex.

As discussed in greater detail below, Zeki and associates (1992) studied each of these visual areas to create a map of the retina. Parallel pathways process various aspects of visual information--color (wavelength), motion, stereopsis, and form (line orientation)--and extend from the retina to the lateral geniculate nucleus, striate cortex, and finally, extrastriate cortex.

Ocular dominance Columns. Ocular dominance columns are seen in the striate cortex. They are seen as an alternating series of parallel stripes that represent a column of neurons in the striate cortex innervated by either the ipsilateral or contralateral eye. Ocular dominance columns extend in alternating bands through all cortical areas and layers, and are absent in only the cortical region representing the blind spot and the cortical area representing the monocular temporal crescent of the visual field. The mosaic appearance of these ocular dominance columns is demonstrated with autoradiography using 2-deoxyglucose and stimulation of only one eye. Simple cells are driven monocularly while the

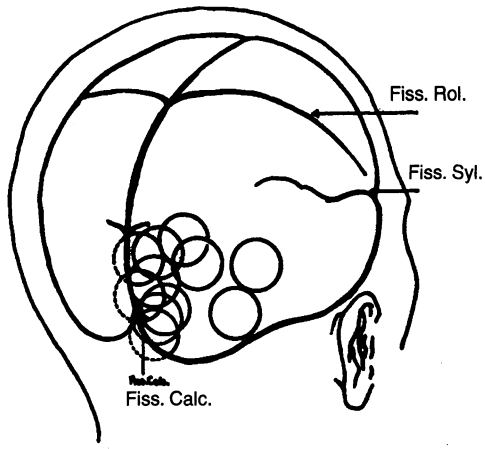


Figure 23-12 Focal discharge in occipital lobe. Location of the lesions at surgery in 11 patients who had focal seizures beginning with visual sensations. (From Penfield, W., and Kristiansen, K.: Epileptic Seizure Patterns, Springfield, Charles C. Thomas, 1951, p.47.)

complex and hyper complex cells are stimulated from both eyes.

Stimulation of Areas 17, 18, and 19

Direct electrical stimulation of areas 17, 18, and 19 in conscious people produces visual sensations (Pollen, 1975) *(Fig. 23-12)*.

These images are not elaborate hallucinations (as in complex partial temporal seizures) but rather are described as flickering lights, stars, lines, spots, and so forth. Often the images are described as colored or moving around. The images are usually localized to the contralateral field, at times to the contralateral eye. An illustrative case, 23-4, is presented below.

At times, the patient cannot determine laterality. In addition, stimulation of areas 18 and 19 (and sometimes 17) produces conjugate deviation of the eyes to the contralateral field and, at times, vertical conjugate movements. Discharge of the occipital cortex may be followed by transient visual defects similar to the transient post-ictal hemiparesis that may follow focal seizures beginning in the motor cortex (see Aldrich et al., 1989). More complete discussion of seizures beginning in occipital cortex can be found in Salanova et al., 1993, and Williamson et al., 1992.

Physiology

The optic (geniculocalcarine) radiation terminates on the cells in the calcarine cortex, which respond like retinal ganglion cells. Their receptive fields consist of an excitatory region surrounded by an inhibitory region. Fields with the opposite pattern are also seen in higher mammals. These fields are excited by an annulus (donut) of light.

Hubel and Wiesel have described three types of higher-order cells in the visual cortex:

1.Simple cell.

2.Complex cell and,

3.Hypercomplex cell.

Simple cell. This cell type responds best to a bar of light with a critical orientation and location. The receptive fields of several simple cells are shown in Figure 23-13. The excitatory field of each simple cell is bordered on one or both sides by an inhibitory field as shown for simple cell a. The small portion of the visual field outlined in Figure 23-13 drives thousands of simple cells, each with a discrete location and orientation. Each simple cell appears to receive input from a number of ganglion cells, which have their excitatory and inhibitory fields in a straight line.

Every simple cell can be driven by input from either eye, but they usually show a preference for one eye or the other. For example, a simple cell may be strongly excited by a bar seen with the right eye and only weakly stimulated by the same bar seen with the left eye. This differential sensitivity may form the basis of binocular vision.

Complex Cell. A number of simple cells having the same orientation activate a complex cell. The complex cell has a definite orientational preference, but a much larger receptive field. An example of this is complex cell alpha in Figure 23-13. Any horizontal line of any length within the outlined visual field excites complex cell alpha. Lines with different orientations in the same visual field will drive other complex cells.

Hyper complex cell. The final cell type is a

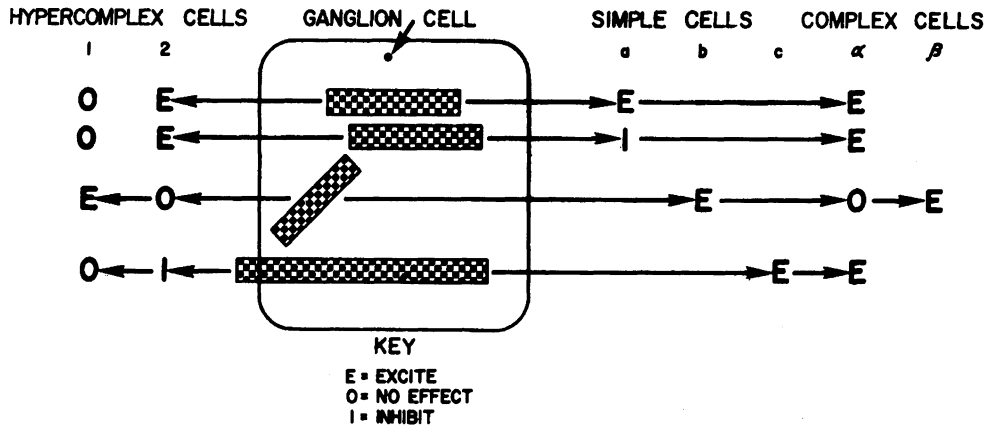


Figure 23-13. Types of visual stimuli that excite each of the four types of cells found in the calcarine cortex. The square represents a box 2 inch by 2 inch, 5 feet away from the monkey's eye. The actual stimuli are bright bars of light on a dark field. (Data from Hubel and Wiesel. 1968. J. Physiol. 195:215.)

hyper complex cell. It has characteristics very similar to the complex cell except it discriminates lines of different lengths. The bottom light bar in *Figure 23-13* will inhibit hyper complex cell 2. If its left-hand border were moved into the visual field, it would strongly excite hyper complex cell 2. Once again, it is excited by lines of a critical orientation anywhere within a large portion of a visual field, but they must not be too long. Corners and angles also excite these cells.

About half a million optic-tract fibers enter each occipital lobe. It should be clear that there are many more cells processing this information, and indeed, the banks of the calcarine cortex contain many millions of cells.

The response to a large rectangle of light varies with the type of cell. Only if the edge falls on the center of a cell's receptive field will it be excited. Ganglion cells are very sensitive to position, contrast, and dynamics. Ganglion cells, in turn, excite simple cells that outline the edges of the rectangle of light. Simple cells have a larger receptive field than ganglion cells, are less sensitive to position, but are sensitive to the direction of the edge. Simple cells do not respond to continuous light or dark stimuli in their fields or to bars of light with an orientation other than their preferred orientation. Complex cells have much larger receptive fields than simple cells but are heavily influenced by the direction of the edge. Lastly, a hyper complex cell must have a corner, an angle, or the end of the edge within its field to be activated.

When a rectangle of light is flashed upon the retina, the cells described are initially activated but adapt fairly quickly and then are hardly stimulated. However, if the pattern flashes on and off, the cells are repeatedly stimulated. The cortex is also stimulated if the pattern moves a little on the retina. A different group of ganglion and simple cells will react, but because of their larger receptive fields, most of the complex and hyper complex cells continue to be stimulated. The nervous system has overcome the problem of stationary images by continually producing rapid, saccadic eye movements that constantly shift the image on the retina. Indeed, when the image is stabilized by special contact lenses, it disappears.

In the process of data reduction, the visual system loses the ability to judge absolute light intensity for the most part.

Differences in intensity within a field can be determined easily but not the overall intensity of the field; photographers who estimate exposures usually waste a lot of film. Only the pupillary reflex system retains the ability to measure absolute intensity.

Different calcarine cells within a narrow

column perpendicular to the surface repeat this processing for each of the primary-color receptors to give information on form, color, motion, and contrast. Lesions as small as 1 mm result in a detectable loss of all of these modalities in a small area of the visual field.

Zeki and associates (1992) demonstrated how the four identified perceptual pathways *(color, form with color, dynamic form, and motion)* relate to the specific visual areas.

The neurons of area V5 (area 19) are responsive to *motion and directionally selective,* but they are nonselective for color. In contrast, the majority of cells in V4 (area 19) are selective for specific wavelengths of light (color sensitive), and many are selective for form (line orientation) as well.

The cells of the adjacent areas V3 and V3A (area 18) are selective for *form (line orientation) but are indifferent to wavelength (color).* V1 (area 17) and V2 (area 18) distribute information to these specialized fields. V1 has column and intercolumnar areas. The columns stain heavily for the energy-related enzyme cytochrome oxidase. The neurons within the columns are wavelength sensitive, tend to concentrate in layers 2 and 3, and receive input primarily from the parvocellular layers of the lateral geniculate nucleus. Information concerning color is then projected either directly or through thin-column stripes in area V2 to V4.

By contrast, *form-selective neurons* are located in the intercolumnar areas in V1. Form-related-to-color derives from the parvocellular-neuron layers of the lateral geniculate nucleus, which project first to the intercolumnar areas of V1 and then to V4.

The second *form system is independent of color* and depends on inputs from the magnocellular layers of the lateral geniculate nucleus to layer 4B of V1 projecting to V3. Extrastriate areas send projections back to the dorsal lateral geniculate nucleus and the pulvinar of the thalamus.

The *motion-sensitive system* consists of inputs from the magnocellular layers of lateral geniculate nucleus to layer 4B of area V1, which then projects to area V5, either directly or through area V2. Each of the prestriate areas V5, V4, and V3 sends information back to V1 and V2, as well as to the parietal and temporal areas. This provides for more integrated visual perception. Striate

To summarize, let us consider the example of a slowly moving colored ball. Visual information is fractionated into the modalities of form, color, and motion in the calcarine cortex. That information is reassembled elsewhere to give us a unitary view of the ball.

We are just beginning to understand where and how this parallel processing is done.

If the object is the letter A, then further processing takes place in the dominant (usually left) lateral occipital cortex (Brodmann's areas 18 and 19), the visual language association area. Letters seen in the left visual field and represented in the right calcarine cortex must project across the posterior corpus callosum before they can be recognized in the left visual association cortex.

Information needed to reconstruct complex forms flows through the inferior occipital/temporal region, primarily on the nondominant side. Located here are cells that can discriminate between human faces, for example. Similarly, sheep have cells that respond only to the image of a wolf (not of a sheep), and monkeys have cells that respond only to a human face but not to a snake or a banana. Many of these cells are sensitive to gaze, being strongly stimulated by direct eye contact but only weakly stimulated when the eyes are averted.

Extra-occipital higher-level visual processing then occurs in area 7 of the parietal lobe, areas 23 and 21 of the temporal lobe, and area 8 of the frontal lobe (see Pandya and Kuypers, 1969).

Occipital Lobe and Eye Movements.

Areas 18 and 19 send fibers to the tectal area of the superior colliculus. Such connections are necessary for visual fixation and accurate following of a moving object. The slow and smooth conjugate eye movements that

occur when the eyes are following a moving visual stimulus (pursuit movements) should be distinguished from the independent phenomenon of voluntary (saccadic) conjugate eye movements.

Saccadic movements are rapid and shorter in latency and duration. They do not require a visual stimulus and do not depend on any connections to area 18. Rather, they are dependent on area 8, which sends fibers to the lateral gaze center of the pons.

This discussion of cortical control of eye movements is clearly simplified (see also chapter 18). Both areas send some fibers to the superior colliculus, and ultimately, both the saccadic and pursuit movements involve the pontine gaze centers. In addition, vestibular and cerebellar influences also determine eye movement.

Lesion in Area 17

Complete unilateral ablation of area 17 (V1) produces a complete homonymous hemianopia in the contralateral visual field (the same half field in each eye has no conscious visual perception). Such patients may have some crude visual function such as spatial localization, presumably because of optic-tract connections to the superior colliculus. A report by Barbur and colleagues (1993) suggests that conscious perception of the type of visual stimulus and its direction of motion can continue to occur after selective V1 lesions have been made, due to activation of V5 (possibly by subcortical input to V5). Vision is clearly abnormal in this blind field. V1 to V5 input is the dominant input in intact individuals.

With partial lesions, a partial defect results. For example, if only the superior bank of the calcarine fissure is involved, the visual field defect is limited to the inferior quadrant of the contralateral field. A bilateral infarct of either the upper or lower bank of the calcarine fissure may produce a homonymous altitudinal hemianopia in which apparent blindness exists in either the entire lower or upper field of vision (as appropriate).

With such calcarine-cortex lesions, the field defects are usually similar (congruous) in both eyes. By contrast, incomplete optic-tract lesions may produce somewhat unequal (non congruous) homonymous field defects in both eyes.

Vascular lesions, in particular occlusion of the posterior cerebral artery, often result in a homonymous hemianopia with macular sparing. That is, vision in the macular area of the involved field remains intact. Such preservation of macular vision probably occurs because the macular area has a large representation in the most posterior third of the calcarine cortex and is the area nearest the occipital pole. This area, then, is best situated to receive leptomeningeal anastomotic blood supply from the middle cerebral and anterior cerebral arteries. Occasionally, with occipital infarcts, there may be preservation of vision in a small peripheral unpaired portion of the visual field, called the temporal crescent (see Benton et al., 1980).

In vascular insufficiency or occlusion of the basilar artery, infarction may occur in the distribution of both posterior cerebral arteries, producing a bilateral homonymous hemianopia and a syndrome of "cortical blindness." Such patients lose all visual sensation, but pupillary constriction in response to light is preserved.

Lesions of Extrastriate Areas 18 and 19

Lesions in areas 18 and 19 often referred to as the visual association areas, produce deficits in visual association, including defects in visual recognition and reading. The problem is that in humans with a unilateral lesion, one is almost never dealing with disease limited to areas 18 and 19 (however, see discussion below of more recent studies).

Rather, one finds that adjacent portions of either the inferior temporal areas or of the inferior parietal lobule (the angular and supramarginal gyri) are involved or that the lesion extends into the deeper white matter of the lateral occipital area and involves association and callosal fibers. Such more-extensive

lesions will, of course, produce the deficits previously noted in the discussion of parietal function. A limited unilateral ablation might produce defects in visual following, as tested by evoking opticokinetic nystagmus (e.g., moving vertical black lines on a white background or looking at telephone poles from a moving train).

Bilateral lesions limited to areas 18 and 19 occur only rarely in humans. In humans such a lesion would deprive the speech areas of the dominant hemispheres of all visual information. The patient would presumably see objects but be unable to recognize them or to place visual sensations in the context of previous experience. The patient would be, moreover, unable to relate these visual stimuli to tactile and auditory stimuli. In monkeys, restricted lesions in the "visual" areas of the temporal and parietal lobes also interfere with pattern discrimination (Mishkin, 1972).

Horton and Hoyt (1991) have reported that unilateral lesions specific to the extrastriate V2/V3 cortex produce quadrantal visual field defects. As discussed by Zeki (1992), rare patients with restricted lesions in V4 present with achromatopia. The patients see the world only in shades of gray but perception of form, depth, and motion are intact. Rare patients with lesions limited to V5 have akinetopia.

Stationary objects are perceived, but if they are in motion, the objects appear to vanish. Plant and coworkers (1993) have reviewed such impaired motion perception. Selective deficits in form perception are even less frequent. Destruction of both form systems and therefore of V3 and V4 (areas 18 and 19) would be required and, as discussed above, such a lesion would also destroy V1 (area 17), resulting in total blindness.

The following case illustrates the effects of a space-occupying lesion in the occipital lobe. One should compare these findings to those reported earlier in Case 23-1, a lesion in the visual system anterior to the optic chiasm.

Case History 23-3 *(Fig 23-14)* This 47-year-old white, right-handed, real estate salesman developed a cough with some blood present in the sputum (hemoptysis), one month before admission. Five days before admission the patient developed a generalized headache, which was precipitated by coughing or straining and which awakened him or prevented him from sleeping. Two days before admission, the patient noted blurring in the left inferior quadrant of his field of vision. The day before admission, he noted complete loss of vision in this quadrant. On the day of admission, the headache increased, and the patient was unable to see anything in the left visual field. Past history was significant. The patient had multiple pulmonary infections, treated with antibiotics.

Neurologic examination: *Cranial nerves:* Early papilledema was present. A noncongruous left homonymous hemianopia was present *(Fig. 23-14A)*.

Clinical diagnosis: Mass lesion right occipital? Tumor, abscess.

Laboratory data: *Imaging* demonstrated an enhanced lesion in the posterior and medial aspect of the right hemisphere (occipital and adjacent parietal lobes). *EEG* was abnormal because of frequent focal 3 to 4 cps, slow waves in the right occipital area and to a lesser degree, the right posterior parietal area. *Cerebrospinal fluid* pressure was elevated to 210 mm CSF, 97 lymphocytes were present (upper limit is 5 to 7 lymphocytes). Protein was increased to 75 mg/dl. Glucose was 75 mg/dl (normal when compared to blood sugar of 95 mg/dl).

Subsequent course: The patient was treated with antibiotics (cephalothin sodium, penicillin, and streptomycin), and visual fields, EEG, and spinal fluid findings improved. Within 3 weeks the field defect had resolved to a non congruous left inferior quadrantanopia *(Fig. 23-14B)*.

Ten days later the patient was readmitted to the hospital with a three-day history of right eye pain, sweats, and chills. Neurologic examination now revealed a recurrence of blurred optic-disc margins, a left homonymous hemi-

anopia and a slight increase in deep tendon reflexes on the left. Imaging studies now revealed a large space-occupying lesion in the right parietal/occipital region, displacing the right lateral ventricle forward and downward. After 10 days treatment with antibiotics (penicillin and streptomycin), a craniotomy was performed by Dr. Bertram Selverstone revealing an abscess and a large surrounding area of hard granulomatous cortex and white matter that were removed. The etiologic organism was subsequently found to be a microaerophilic streptococcus.

Follow-up examination 6 months after surgery was normal, except for the left homonymous hemianopia *(Fig. 23-14C)*.

The case that follows is another example of the effects of a lesion in the occipital lobe with very different consequences then those seen in the previous case.

Case History 23-4. This 18-year-old left-handed single white female restaurant employee had the onset of her seizures at age 14 when she had a sequence of five seizures in less than 12 hours. Each began with flashing lights, "like Christmas tree lights," all over her visual field, plus the sensation" that peoples' faces were moving. She then would have an apparent generalized convulsive seizure. Neurologic examination, and CT scan were all reported as normal. An electroencephalogram reported occasional focal sharp and slow waves in the left hemisphere. The patient was treated with carbamazepine (Tegretol), 400 mg in the morning, and 230 mg at hour of sleep with no additional

She apparently had done quite well with no additional grand mal seizures. She had rare episodes of "fear attacks," which would last 23 to 25 minutes. The last attack had occurred 2 years ago, but as recently as two months ago, she had had one episode of flashing lights. A recent EEG was normal.

Neurologic examination: normal.

Clinical diagnosis: Seizures of focal origin left occipital.

Subsequent course: The patient did well

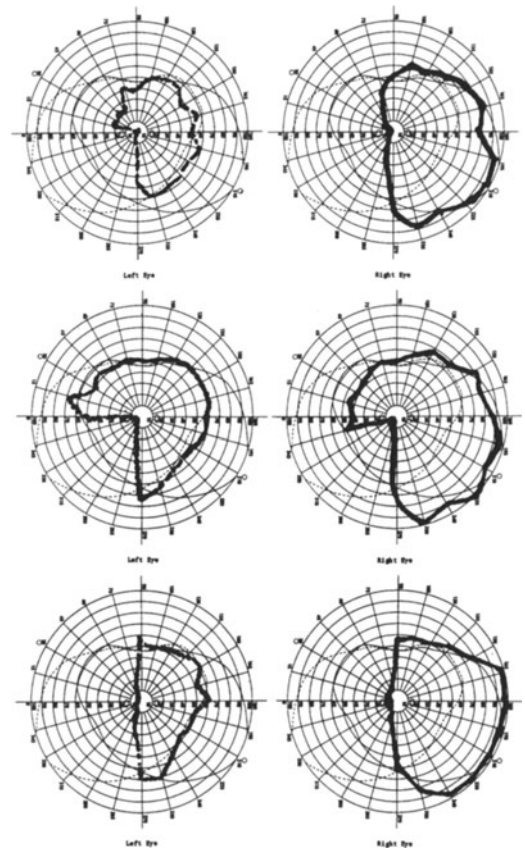


Figure 23-14. Case History 23-3, Brain abscess, right occipital area: Perimetric examination of visual fields. A, Initial examination demonstrated a somewhat asymmetrical (non congruous) left homonymous hemianopia less in the left eye than the right. The fields are shown from the patient's point of view. B, 16 days later; with antibiotic therapy, an improvement had occurred. A non congruous, quadrantanopia is now present. C, Fields after an additional 24 days. A relatively complete homonymous hemianopia, present at the time of readmission, persisted following surgery as shown above.

for 3 years then had a recurrence of a generalized convulsive seizure possibly related to omission of medication. Six weeks later, she reported two additional episodes characterized by flashing lights, then movement of the lights away from a center circle, then dizziness, then a sensation of unreality. She also had other episodes of feeling unreal accompanied by fear.

The neurologic examination demonstrated a minor right central facial weakness not present previously.

Laboratory data: *EEG:* normal. *MRI*

scan now revealed a small tumor at the left occipital pole with surrounding edema *(Fig. 23-15)*. *CT scan*, this tumor appeared partially calcified but did show enhancement. Review of the CT scan obtained in Arizona at age 14 indicated similar findings. Angiograms suggested tumor vascularity of a type seen with meningiomas.

Dr. Bernard Stone removed a discrete encapsulated tumor that appeared to be a meningioma. Subsequent microscopic examination, however, indicated a rare type of indolent follicular adenocarcinoma of the thyroid, which sometimes spreads as a single lesion to brain and remains quiescent for many years. In the postoperative period, a non-congruous right-inferior-field defect was present (partial quadrantanopia). The blood level of thyroid-stimulating hormone (TSH) was elevated. A thyroid nodule was found. A thyroidectomy was performed, and thyroid replacement medication was prescribed. No additional seizures were observed over the next 18 months. The patient subsequently developed a lumbar vertebral metastasis.

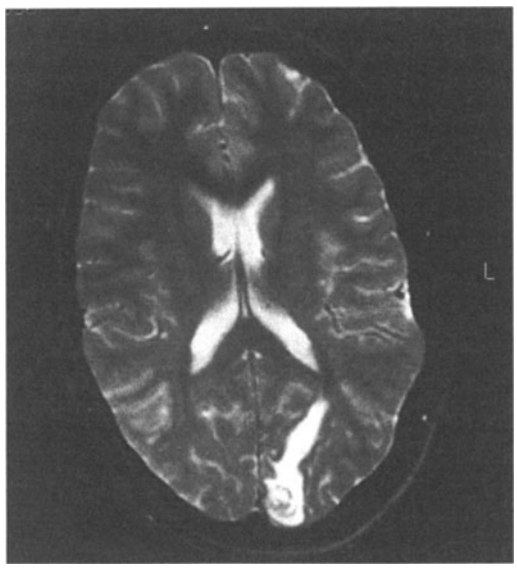


Figure 23-15 Case 23-2. Focal visual seizures characterized by "flashing lights" and the sensation of movement of the visual field with secondary generalization beginning at age 14 due to metastatic (thyroid) tumor of the left occipital lobe. MRI, T2-weighted, non-enhanced demonstrated a small tumor at the left occipital pole with surrounding edema. (See text for details.)

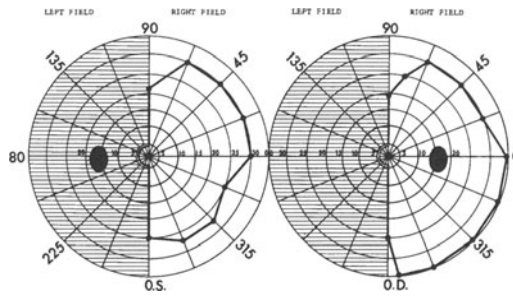


Figure 23-16. Occlusion of right posterior cerebral artery. Case 26-6 on CD ROM. Tangent screen examination of visual fields, 3 months after the acute event. This 75-year-old woman had the acute onset of headache, bilateral blindness, confusion, vomiting, mild ataxia and bilateral Babinski signs. All findings cleared except a residual left homonymous hemianopsia with macular sparing. O.S. = left eye; O.D. = right eye.

Vascular lesions of the calcarine cortex: Various types of visual field defects may occur including a homonymous hemianopsia, with *(23-16)* or without macular sparing, or a quadrantanopsia. The CT scan correlations are presented in *Figures 23-17 and 23-18.*

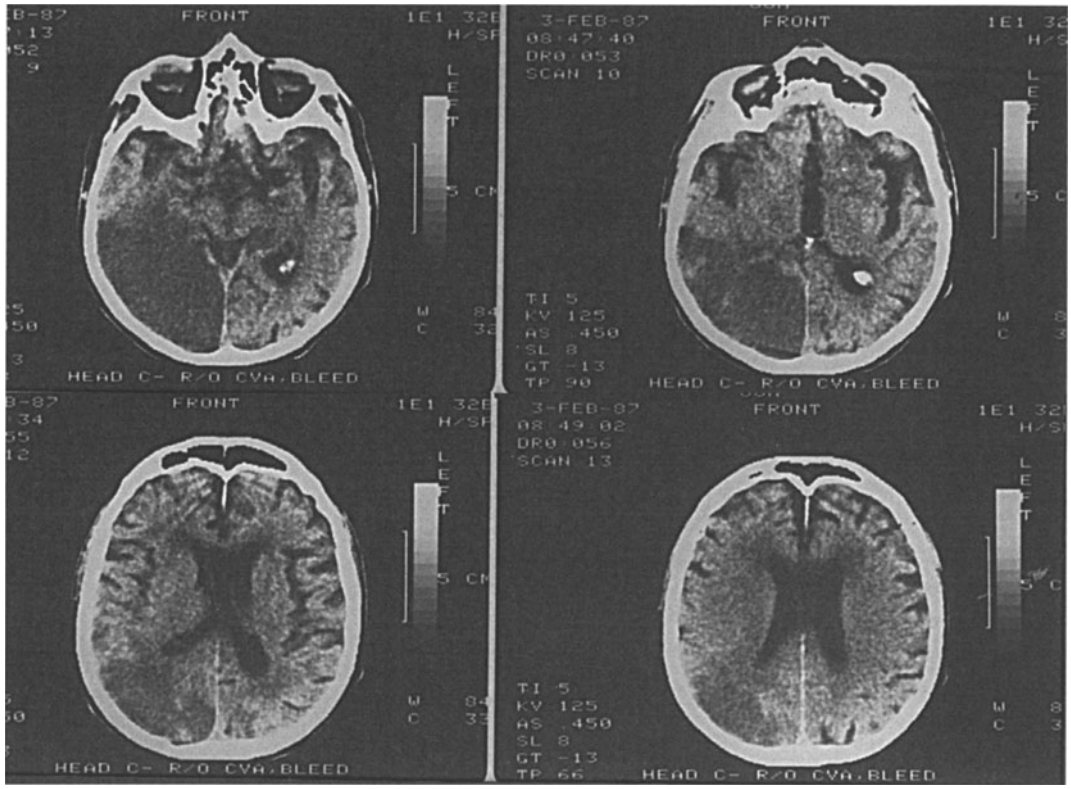


Figure 23-17 CT scan of a total infarct, presumably embolic in the right posterior cerebral artery cortical territory (occipital and posterior temporal lobes) obtained 5 days after the acute onset of confusion and possible visual hallucinations in an 86-year-old right-handed male. As confusion cleared examination indicated a dense left homonymous hemianopia with no evidence of macular sparing. The patient would look to the right and to the midline but would not follow objects to the left beyond the midline. Contrast to Fig. 23-18.

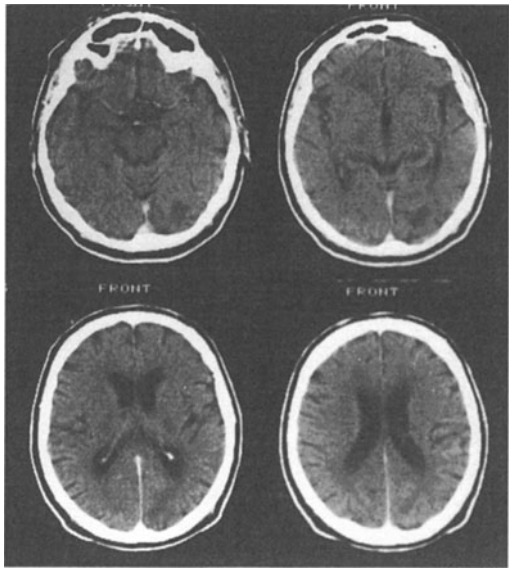


Figure 23-18. CT scans of a partial infarct of the left posterior cerebral artery territory (inferior calcarine) with superior quadrantanopia probable basilar vertebral ischemia with artery-to-artery embolic infarction of the occipital (calcarine artery). This 71-year-old right-handed male with a history of diabetes mellitus and hypertension had acute onset of vomiting, ataxia, minor confusion, and a persistent problem with peripheral vision in the right visual field, followed by episodes of vertical diplopia. Examination the following month demonstrated a homonymous visual field defect involving the right superior field, minor ataxia, and a right Babinski sign. Compare to figure 23-17.