

Functional Aspects of Neural Visual Disorders of the Eye and Brain

14

ARIES ARDITI, PH.D.
JOSEF ZIHL, PH.D.

From the discrete sampling of the optical image by the photoreceptor mosaic to subsequent cortical neural image processing and cognition, visual perception is comprised of a wide range of transformations and relations. Their main common property is that they arise from light stimulation of the retinal photoreceptors. This chapter is a brief introduction to how age-related disruptions of that neural structure degrade visual perception and lead to low vision in middle and later life. A similar treatment for pediatric low vision is in the chapter by Hartmann (Chapter 12).

Historically, the clinical subspecialty of low vision grew out of ophthalmology and optometry. As a result, the field has tended to focus on dysfunctions of the eye and less so on those of the visual areas of the brain, which have traditionally been viewed as the domain of the neurologist. This chapter attempts to present a simple, unified view of neural disorders leading to low vision, with respect to locus in the visual pathway of the disorder. It views neural visual disorders of eye and brain alike, as anomalies or interruptions in visual processing streams. This chapter also attempts to relate these to visual functions that are assessable through psychophysical or neuropsychological testing.

Myriad disorders and diseases can destroy neural tissue of the visual system in the retinas, optic

nerves, and brain. Mechanical forces, ischemia, anoxia, infection, tumor, degenerative processes, and hemorrhage are among the many prior causes that may lead to photoreceptor or other nerve cell losses anywhere along the visual pathway. In all these situations, the end result is nerve cell death and interruption of the nerve signal transmissions that collectively comprise the visual sense. Rather than focus on such causes, the chapter examines how disrupted anatomic and physiological functions produce deficits in measurable visual functions.

Many common optical disorders of the eye occur in middle and later life, as described in Chapter 13 of the handbook. These may coexist with neural disorders of the eye and brain. Ocular media opacities and reduced pupil size (miosis) are typical changes in older adults. These reduce intensity and, in the case of opacities, overall quality of the retinal image and wavelength composition of light entering the eye. Where such conditions coexist with neural conditions, the perceptual consequences for the patient may be compounded.

Basic Concepts

Figure 14.1 illustrates the gross neuroanatomy of the visual system. A basic knowledge of the paths of fibers from each half of the retina to the striate cortex, where much early visual processing takes

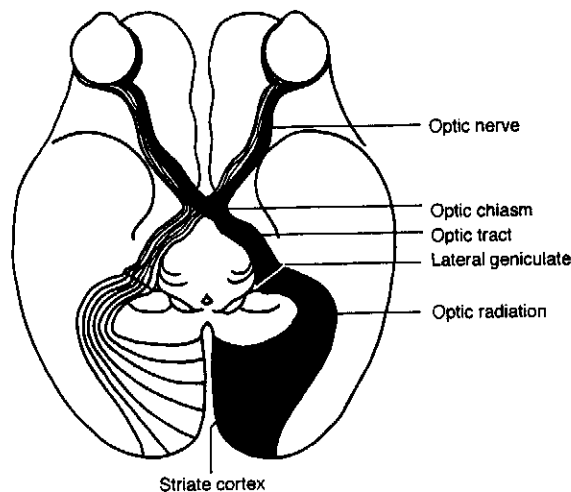


Figure 14.1. The optic nerve decussates at the optic chiasm so that nasal optic nerve fibers (serving the temporal visual field) travel to the contralateral side of the brain; temporal optic nerve fibers (serving the nasal visual field) travel to the ipsilateral side of the brain. This arrangement means that damage to the eye or to the optic nerve up to the chiasm will affect vision in one eye or the other, whereas damage beyond the chiasm will tend to affect vision on the contralateral hemifield in both eyes.

place, is essential in understanding which portions of the two eyes' visual fields are affected by neural visual disorders. The following is an overview of the fundamentals of the relationship between the visual fields and the anatomy of the visual pathways. It also includes an introduction to some basic terminology.

The visual field is the set of angular spatial locations to which an eye visually responds when a person's gaze is held in a constant position. Because the images on the two retinas are inverted, light impinging on the temporal retina arises from points in the nasal half of the visual field (that is, hemifield) and vice versa. Thus, in the right eye, the left hemifield is served by the right (temporal) half of the retina. Retinal ganglion cell fibers leave the retinas and carry information to the brain via the optic nerves. The two optic nerves meet at the optic chiasm, where fibers carrying information from the nasal half of the retina decussate and become aggregated with fibers from the temporal retina of the other eye. Posterior to this crossover, the pathway is known as the optic tract. Optic tract fibers terminate in a subcortical region known as the lateral

geniculate nucleus. From there, second-stage neurons carry visual information via the optic radiations to the primary (striate) visual cortex located in the occipital lobe.

If a disease or disorder disrupts neural processing peripheral (that is, anterior) to the optic chiasm (in the retina or optic nerve), the deficit is called *prechiasmatic*. A single prechiasmatic defect affects vision in only one eye. If the defect disrupts processing beyond the chiasm (*postchiasmatic deficit*), it will likely affect the visual fields of both eyes (since the optic tract carries information from both eyes), although not necessarily in exactly the same locations in the two eyes. Such defects produce blindness to the same half (left or right) or same quadrant (for example, left superior) in the two visual fields. The defect is said to be *homonymous*. Blindness to half the visual field is known as *hemianopia* (also *hemianopsia*). Blindness to a quadrant of the visual field is known as *quadrantanopia* (also *quadrantanopsia*). Unilateral lesions in the visual pathway from the optic tract to striate cortex produce both these types of homonymous defects. Other patterns of homonymous field defects also occur, depending on the extent of the lesion. Homonymous defects may be congruous (that is, share the same location and shape in the two eyes). They may also be incongruous, where shape but not gross location may differ. Often, with homonymous hemianopia, a central portion of the central visual field is left intact, despite blindness of the rest of the hemifield. This is often referred to as *macular sparing*. It is believed to be due to overlapping blood supply to this functionally important region. This chapter refers to it as *visual field sparing*.

Hemianopia may also occur on the opposite sides of the visual field in the two eyes, where the left field of the right eye and the right field of the left eye are dysfunctional. This defect is known as *bitemporal hemianopia*. It is a *heteronymous* defect since the defect occurs in different hemifields in the two eyes. This pattern of field defect occurs with lesions of the decussating fibers of the optic chiasm, which are located medially within the chiasm. Such lesions are most commonly due to pituitary tumors that compress these overlying fibers as the tumor expands upward. A lesion in the lateral optic chiasm, of course, can produce blindness to the nasal hemifield of only one eye. Like more peripheral optic nerve

and retinal defects, this deficit is monocular: the deficit exists in one eye alone.

Binocular deficits are those that exist in visual fields of both eyes in corresponding locations. Binocular deficits arise commonly in postchiasmatic injury. However, they are also extremely common in eye disorders because such disorders tend to affect similar field locations in the two eyes, albeit independently and often quite differently in time course and severity. In this context, the term *binocular* simply refers to the corresponding location of bilateral defects. It does not refer to binocular visual functions such as coordinated eye movements, stereoscopic depth, and resolution of diplopia (single vision). Another potential source of confusion is that in prechiasmatic disorders, the term *bilateral* refers to the presence of the condition in the two eyes (but not necessarily in corresponding locations). In postchiasmatic injuries, however, it refers to both the left and right hemispheres of the brain or, correspondingly, to the left and right sides of the visual field.

Visual field maps, which provide the richest source of information about visual function in postchiasmatic defects and in some prechiasmatic defects, are most effectively mapped with a perimeter. This is a device that presents stationary or moving points of light against a white background of standard luminance. The patient, who maintains his or her gaze on a fixation point, reports on the visibility of the lights by means of a button press. These responses are recorded on a standard polar coordinate visual field chart. The many sorts of visual field maps summarize which (and often the extent to which) visual field locations, and hence the retinal regions that serve them, are functional. Figure 14.2 shows the extent of the normal visual field and the extent of locations corresponding to the central retina (conventionally ± 15 degrees surrounding the foveal origin), the macula, the fovea, and the rod-free foveola. In individuals of European descent, the normal field is usually limited in extent nasally and superiorly by the nose and brow, respectively.

Perimetry is most useful for assessing neural visual disorders that produce blindness or reduced function in a significant portion of the visual field. However, it is of limited value in assessing disorders in the central 10 degrees of vision. This is so for several reasons. First, scotomas (regions of blindness in the visual field) very small in area often cannot

be detected with perimetry at all. Despite their small size, however, such scotomas in the central 10 degrees of the visual field often significantly diminish visual function since this region supports better visual function (for example, resolution of the neural image, contrast sensitivity, pattern discrimination capability, or color discrimination) than does any other region. In contrast, in the peripheral visual field, even very large scotomas may go unnoticed except when detected with perimetry. Perimetry is simply too crude to yield much useful information about function in the central 10 degrees of the visual field.

The second reason why perimetry is of limited value in this region is that fixing the patient's gaze accurately and consistently during perimetry is difficult. Steady fixation results in a horizontal gaze standard deviation of some 8 min arc in untrained observers who are asked to fixate for only 12 seconds (Kosnik, Fikrer, & Sekuler, 1986). Accuracy will be far worse when observers must fixate for the tens of minutes required for any but the most cursory perimetric evaluations. This makes it difficult to localize field defects with great accuracy. Also, when perimetric stimuli are brought close to the fixation point, patients experience a physiological compulsion to direct the gaze toward the target, further reducing the accuracy of both the fixation and the perimetry. While reduced accuracy affects mapping of peripheral as well as central defects, incremental losses in the central field are, as noted above, vastly more important functionally than those in the peripheral field area. The recently developed confocal scanning laser ophthalmoscope allows direct imaging of the fundus of the eye. With certain modifications, it makes possible video presentation of visual images directly onto the retina while image location is being observed. This technology, currently used only in research settings, holds great promise in increasing the spatial accuracy of central field assessment since perimetry can be performed during direct viewing of the fundus (Sunness, Bressler, & Maguire, 1995).

A third reason why perimetry of the central 10 degrees is problematic is that the presence of neural deficits in and around the fovea makes voluntary fixation extremely difficult for the patient. This occurs since, in the perimeter bowl, the patient has little visual information to guide eye position. Thus, it is fairly well accepted that neither static (inten-

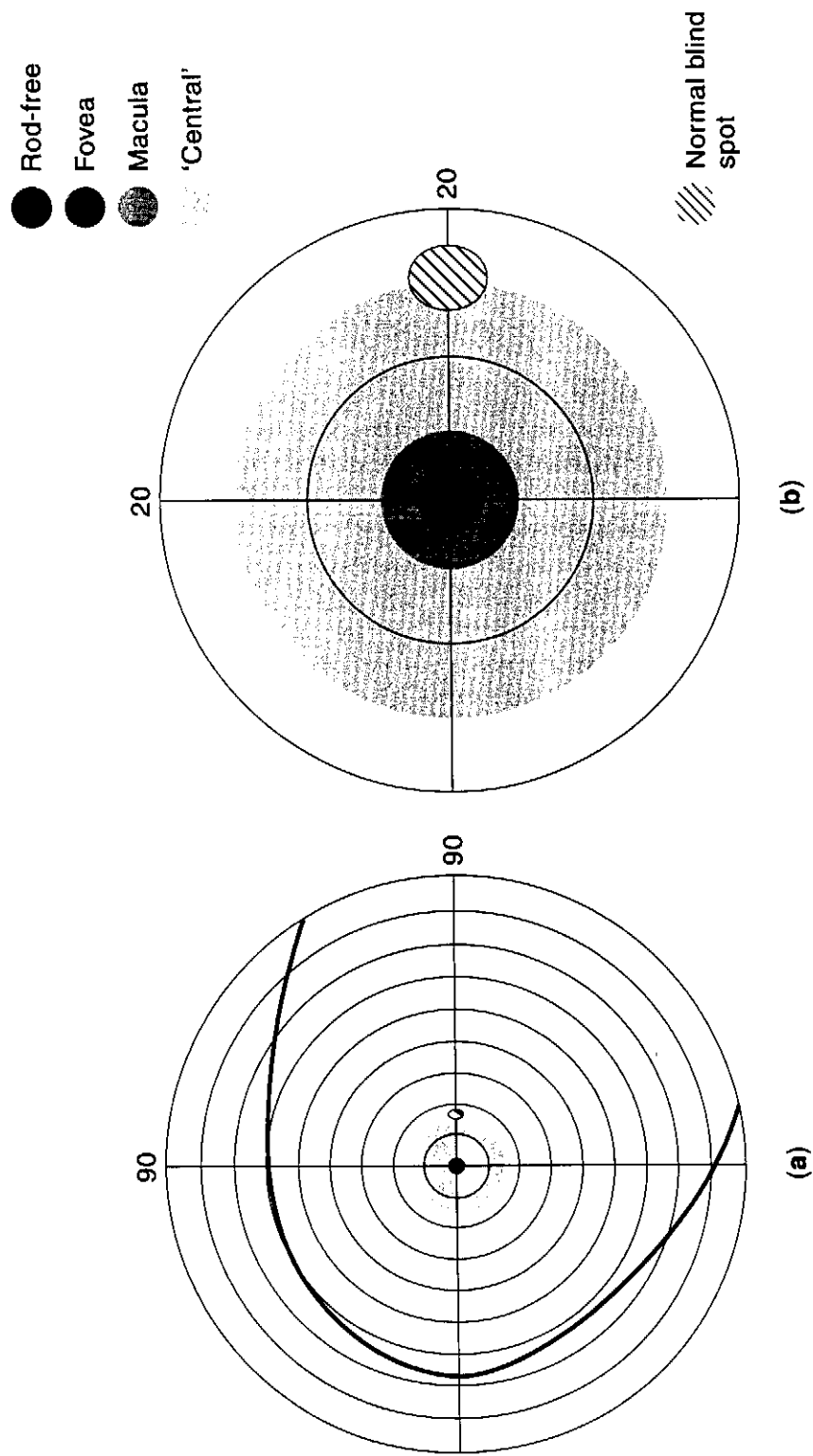


Figure 14.2. (A) The extent of the normal visual field. (B) Exploded view of (A), showing central 40 degrees.

sity thresholds to flashed stimuli) nor kinetic (field-position thresholds of fixed-intensity light moving from periphery to center or vice versa) perimetric methods yield reliable visual field maps for visual function in this region. Generally, the integrity of this region is clinically assessed by visual acuity and Amsler grid (see later discussion) methods. When there are very large scotomas that encompass the entire central 10-degree region, perimetry may be eliminated altogether as an assessment option. Fields are often assessed with so-called confrontation methods, where the examiner uses a hand motion as a stimulus while monitoring the patient's fixation.

The severity of a visual field defect can vary not only in size and spatial location but also in sensitivity to contrast, which in this context is usually referred to as *defect depth* or *density*. Scotomas may be absolute, in which sensitivity is nil. They may be relative, in which sensitivity is reduced relative to sensitivity in the same region of a typical age-matched observer. Figure 14.3 shows this typical "hill of vision." Contrast sensitivity in the normal visual field is highest in the fovea and falls off with increasing retinal eccentricity. Thus, central visual field defects will generally drastically reduce the patient's overall contrast sensitivity. Additional terminology exists to specify the category

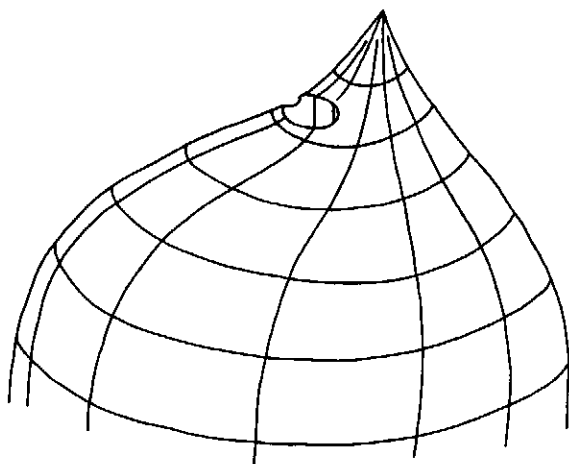


Figure 14.3. Schematic view of the normal hill of vision representing the variation of sensitivity to contrasts as a function of visual field location. Sensitivity is higher at the fovea and falls off monotonically with increasing eccentricity, except in the region of the normal blind spot, where sensitivity is nil.

of defect further (for example, depression, contraction, or scotoma) (Harrington, 1976), but it will be of limited utility in this chapter, which refers to local and extended areas of visual dysfunction as scotomas.

The reader should note that the visual field represents the measured function of one eye alone. Ophthalmologists commonly believe that superimposing the visual fields of the two eyes and evaluating which visual field locations are functional in either eye yield a complete picture of which spatial locations an observer can see. A popular method of functional visual field evaluation has been based upon this assumption (Esterman, 1982). However, the assumption is not valid. The volume visual field (Arditi, 1988), which is the set of locations in space that an observer can see with fixed eye position, simply cannot be represented in fewer than three dimensions. This is because scotomas in the two horizontally disparate eyes serve spatial locations that lie at some distance removed from the fixation plane. For example, in figure 14.4, light that impinges on the normal blind spot of one eye and the retinally disparate central scotoma of the contralateral eye will arise from a volume of space that lies some distance farther from the observer than the fixation point. The superposition of the visual field maps represents function only on the plane corresponding to infinite viewing distance (Figure 14.5).

Of the many individual types of visual disorder described in the medical literature, prechiasmatic disorders (those affecting the retinas and optic nerves) are considerably more prevalent than disorders of higher visual pathways of the brain, such as strokes, tumors, and traumatic injuries (see Chapter 27). Since neural damage to the retina and optic nerve interrupts the input of visual information to all higher areas of the brain, vision may be completely lost in the affected areas of the monocular visual field. But, since the healthy retina in the fellow eye may compensate for monocular scotomas, substantial vision impairment often results only after disease has affected both eyes. In contrast, lesions in the primary (occipital) visual cortex tend to produce homonymous scotomas, which are far more devastating to perceptual function than are monocular deficits. However, generally within the brain, only in the occipital cortex or its afferent pathways does damage result in scotomas. Extraoccipital injury gener-

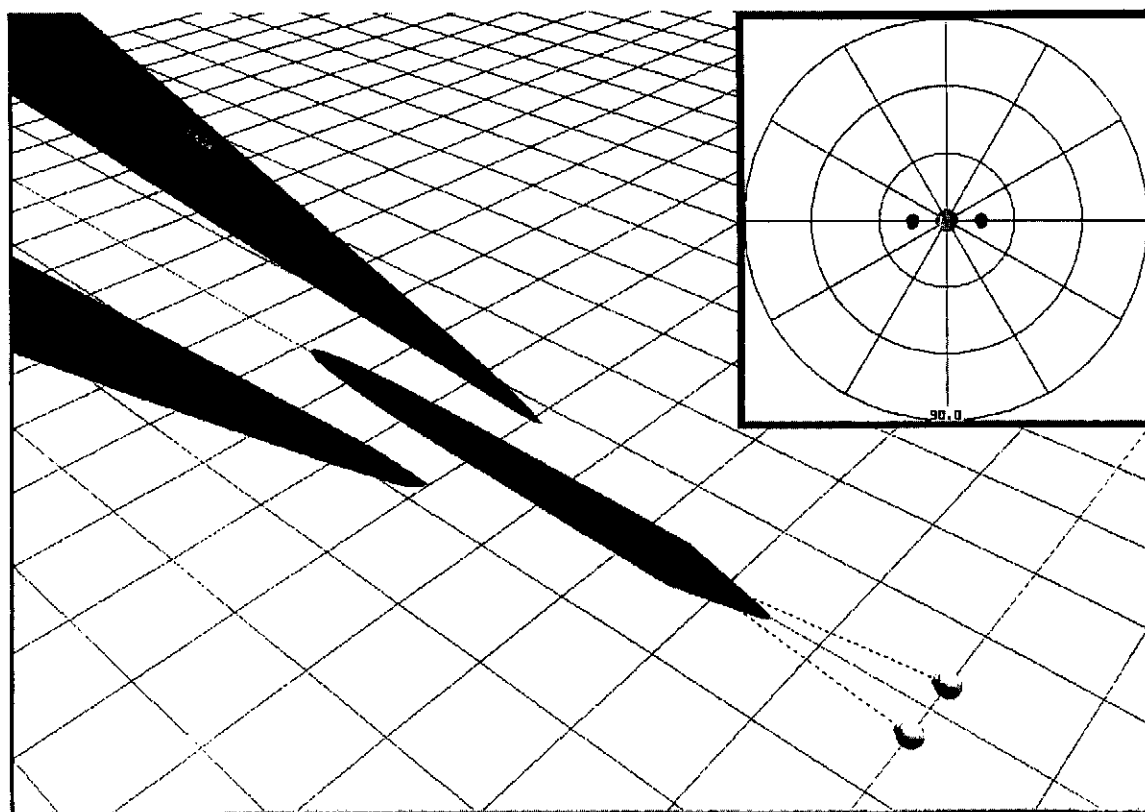


Figure 14.4. The volume visual field of an individual with circular macular scotomas, shown with eyes fixating at the intersection of the dashed lines. Superimposed monocular field maps are shown in the inset, depicting the two normal blind spots and a bilateral central scotoma. This common pattern of visual field defect gives rise to blindness in three distinct volumes of space. One is composed of the intersection of field areas associated with the central scotomas. The other two are composed of the intersection of field areas associated with the central scotoma of one eye with the blind spot of the other eye.

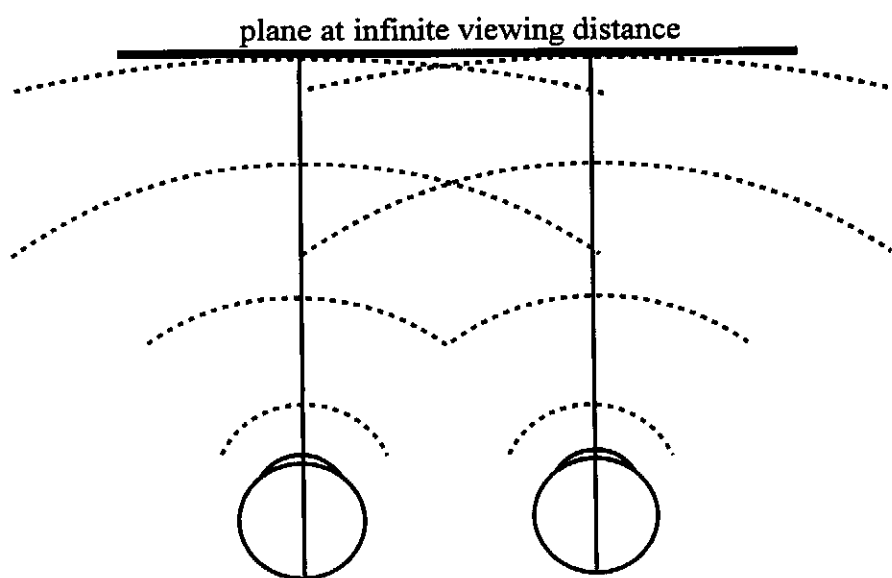


Figure 14.5. The plane represented by the superposition of the visual field maps of the two eyes. This plane can be thought of as a spherical surface, like the inside of a perimeter bowl of infinite size.

ally leaves the visual fields intact and affects only specific perceptual and cognitive functions.

Prechiasmatic Neural Deficits

Visual neural deficits arising from disorders peripheral to the decussation of the optic nerve at the optic chiasm are always monocular deficits unless bilateral involvement occurs. In a sense, this makes these deficits less severe than postchiasmatic occipital deficits since the functional retina in the fellow eye may serve to compensate to some degree. This compensation is true mainly to the extent that the object of visual interest falls on corresponding locations in the two retinas and is thus lying at the same depth plane. However, monocular deficits may adversely affect binocular function. This can lead to diplopia (double vision), disruption of stereoscopic depth perception, and a variety of other visual performance decrements. Monocular deficits, of course, may occur independently in both eyes and in similar or the same (in which case they become binocular deficits) locations in the two visual fields.

Central Monocular Field Defects

Central monocular field defects affect the central 30 degrees surrounding the foveal origin (see Figure 14.2). The most common diseases of middle and later life that affect this region are macular degeneration, chronic open-angle glaucoma, and diabetes. Macular degeneration produces a wide variety of defects anywhere within the macula and especially in the more vulnerable fovea where nutritive requirements are high and the retina thins. Glaucoma tends, in early stages of field loss, to produce arcuate (arc-shaped) defects involving the field around the optic disk and, later, more extended losses in the midperiphery and far periphery. The central few degrees of field are usually intact until the final stages of field loss progression (which can usually be arrested with drug therapy). Finally, diabetic retinopathy, through retinal hemorrhaging, retinal detachment, or photocoagulation treatments, often results in defects in the central visual field (but may also occur in the peripheral field), depending on location of the retinal disease. (Discussion of the medical aspects of these eye diseases is provided in Part I of the handbook.)

Types of Central Monocular Field Defects

Central monocular field defects may be further subdivided into foveal and nonfoveal central defects. Purely foveal defects are confined to the foveal visual field region (though not necessarily of retinal etiology). They are too small to be revealed through perimetry but may affect perception or discrimination of fine details in visual patterns. Such defects may be detected by the Amsler grid, a subjective test in which the patient reports on or draws impressions of distortions of a regularly spaced grid. The only known disorder thought to affect the foveal visual field specifically is amblyopia, a developmental condition with two forms. One is associated with unequal refractive states in the two eyes (anisometropic amblyopia). The other is associated with favoring one eye's image due to extraocular muscle imbalance (strabismic amblyopia). In both forms, the development, in the first years of life, of neural circuits for fine-pattern discrimination appears to be disrupted due to inadequate detail in foveal imagery. While amblyopia is not a disorder of middle and later life, the understanding of foveal function has been greatly enhanced by its study.

In middle and later life, macular disease may diminish or eliminate foveal function. However, as noted, precise perimetric methods do not exist for evaluating the central few degrees of visual field. Most macular defects encompass the fovea but are not confined to it. These defects interfere with both the resolving capacity of the eye and the ability to evaluate relative object positions accurately. In general, the larger the affected central region, the greater the magnification required for tasks involving detail vision, including object recognition and fine localization.

Some monocular defects in the central 30 degrees of the visual field do not engulf the foveal region. Such defects, especially common in glaucoma, may not affect visual resolution. Like the physiological blind spot, the individual may not notice them.

The central 30-degree region is where functional properties of the retina vary most widely in normal vision. The easiest way to understand how a central defect will affect visual performance is to consider how normal functioning would be affected by such a loss. Functional properties and variations of three sorts serve to explain the great majority of the

difficulties experienced by those with central defects: (1) the differences in the response to light by rod and cone photoreceptors, (2) the distribution of these two receptor types across the retina, and (3) the convergence of photoreceptor signals onto retinal ganglion cells.

Rod and Cone Photoreceptor Responses to Light.

The rod receptors are optimized for viewing under conditions of low illumination. However, their responses saturate at light levels relatively low for cones. Rods are so sensitive as to be capable of initiating a visual event in response to even a single photon of light (Baylor, 1987; Hecht, Schlaer, & Pirenne, 1942). Cones, on the other hand, are insensitive to light levels in which rods comfortably convey intensive information. Cones saturate at only the highest levels of environmental illumination. By having three distinct subtypes responsive to different wavelengths of light, the cones are sensitive to and convey information about the spectral properties of light that, after higher processing by the neural pathway, leads to the perception of color.

Figure 14.6 shows the relative sensitivity of the rod and cone systems under conditions that change only the absolute sensitivity of the observer to light (such as changing retinal location of the light stimulus). This depicts the behavior of Hood and Finkelstein's (1986) simplified model in which the two photoreceptor systems are assumed to be independent, spectral sensitivities are assumed to be invariant, and the overall sensitivity of the observer is determined by the most sensitive system for a given stimulus. At low-adapting (scotopic) luminances (Figure 14.6A) in which absolute sensitivity to light is greatest, the rod system determines sensitivity at all wavelengths. At high (photopic) adapting luminances (C), the cones' sensitivity solely determines overall sensitivity. At medium (mesopic) luminances (B), where rod and cone systems may both convey information, the detection is mediated by rods at short wavelengths and by cones at longer wavelengths.

Distribution of Photoreceptor Types. Figure 14.7 shows the distribution of rods and cones across the main horizontal meridian of a retina. Note that the cones are highly concentrated within the central

30 degrees of the retina and are relatively sparse elsewhere. (This coincides with the common definition of the central visual field as 30 degrees in diameter.) The rods, on the other hand, are most numerous at roughly 20-degree eccentricity. They have substantial density everywhere else in the retina, with two exceptions. The first is in the central 1.7 degrees, where only cones exist. The second is in the physiological scotoma (normal blind spot), located 13–18 degrees into the temporal visual field, where there are no receptors at all. This blind area corresponds to the optic disk and is the exit point of the thick bundle of optic nerve fibers from the eye. Thus, the rods are far more numerous than the cones, and the cones populate only the central retina.

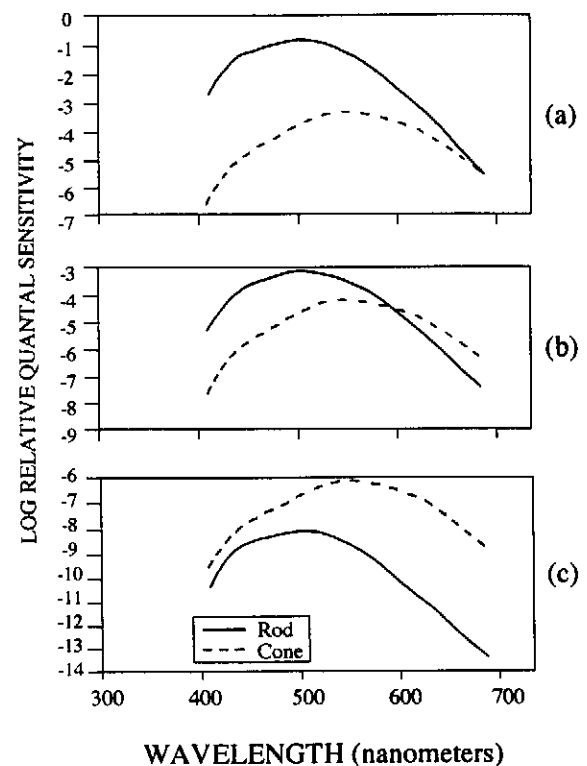


Figure 14.6. Changing relative sensitivity of the rods (solid curve) and cones (dashed curve) to light under conditions that alter the dominance of one system over the other, such as changing adapting luminance or retinal location of the test stimulus. The spectral sensitivity of both rod and cone systems is assumed to be invariant. Changing conditions are assumed to affect only the relative sensitivity of the two systems. (Adapted from Hood and Finkelstein, 1986)

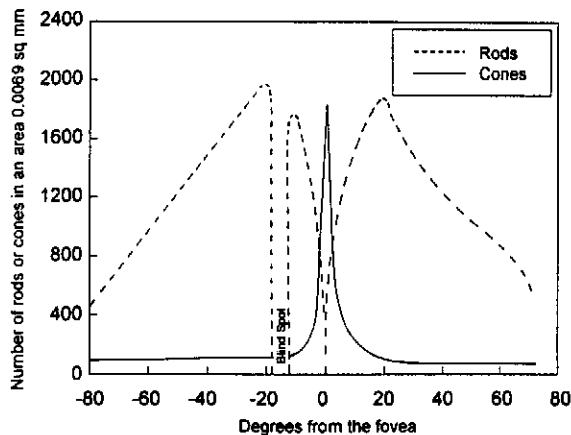


Figure 14.7. Distribution of rods and cones across the main horizontal meridian of the retina.

The dominant role that the small, central 30-degree region plays in photopic vision is reflected in the high kurtosis of the distribution of cones. Beyond 20-degree eccentricity, the number of rods falls off sharply as well, reflecting the dominance of the central visual field in rod vision. From the small proportion of the visual field that central vision represents, it should be clear that a field loss within the central 30 degrees of vision will have a far greater impact on visual function than a loss outside that region.

Convergence of Photoreceptors onto Ganglion Cells. Finally, the convergence ratio of photoreceptor to retinal ganglion cells differs between the two classes of photoreceptor. A great many rods (indirectly) converge on each retinal ganglion cell on the order of 100:1. This convergence allows the ganglion cells to aggregate the responses of rods from a broad retinal region, further enhancing sensitivity but at the expense of visual resolving capacity. Thus, the rod system can detect tiny amounts of light anywhere within a broad region but is poor at discriminating fine detail in patterns due to this pooling of information. The cone system excels at coding the precise location of objects in the visual field because of both its finer photoreceptor spacing and its one-to-one receptor-to-ganglion cell ratio in the fovea. The finer spacing allows resolution of details in the neural image. In contrast, the unity ratio allows the brain to compare magnitudes of light responses of adjacent cones, leading to extremely accurate discrimination of the relative position of retinal image elements.

Effects of Receptor and Optic Nerve Dysfunction on Selected Visual Functions

Given the above picture of normal retinal structure and function, it is possible to understand most of the functional difficulties that people with central visual field defects experience.

Light and Dark Adaptation. The ability to adapt to light and dark is almost always affected when perimacular loss (as often occurs in glaucoma) significantly reduces the number of rods and when central field loss eliminates large numbers of cone photoreceptors. With perimacular loss, not surprisingly, night vision and vision under other low-luminance conditions are especially reduced (see Figure 14.6). When a significant proportion of cones are lost, contrast sensitivity is reduced. More light and higher contrasts are required for adequate pattern vision. In addition, vision is more likely to rely on the rod system, even at photopic light levels. However, since the rods saturate at relatively low light levels, intensity discrimination (and hence pattern vision) is reduced. Patients often attribute their reduced pattern vision under these conditions to glare. Thus, paradoxically, patients with central field loss usually require copious amounts of supplementary illumination but often, at the same time, are glare sensitive, making high illumination uncomfortable.

With both macular and perimacular losses, the range of luminances that the patient can function in is restricted. Furthermore, since the visual system relies on information from both receptor systems to adjust pupil size and control other light and dark adaptation mechanisms, the rod/cone imbalance further reduces the patient's ability to adapt to changes in light intensity.

Visual Resolution. Resolution acuity, often measured with black-and-white gratings of decreasing coarseness, declines with increasing eccentricity in the normal visual field. It does so in a manner that corresponds well with the shape of the decline of cone density with increasing eccentricity. Foveal field defects force the patient to use a more eccentric field location. The fall-off of cone density predicts well the maximum resolution of an individual with a visual field defect involving the fovea.

Visual Localization. Foveal disorders are also associated with reduced ability to localize features in the retinal image as well as to resolve fine details in the image. Localization errors produce reduced vernier, bisection, and stereoacuties—examples of what are now often referred to as the hyperacuties, after Westheimer (1979). Localization deficits may also be accompanied by spatial distortions of the image, or metamorphopsia, as illustrated in Figure 14.8. Such distortions may themselves be seen as a manifestation of the localization errors. Theoretically, cone dropout (that is, death) or physical relocation of cones can explain these localization errors. However, both are difficult to observe in the living eye. Since similar distortions and the position errors are also observed in amblyopia (Levi, 1991), where the retina appears healthy, and in scotomatous regions (Schuchard, 1991), ascertaining whether any given case is due to receptor dropout, relocation, or spatial effects higher in the brain is difficult.

Contrast Sensitivity. Since contrast sensitivity in the normal visual field is markedly higher in the fovea and surrounding macula, central visual field defects will reduce the patient's overall contrast sensitivity. Note that static perimetry itself tests contrast sensitivity to a punctate spot stimulus rather than to a spatially extended grating, which is the more common stimulus used in testing con-

trast sensitivity. Both contrast sensitivity testing and perimetry generally use photopic luminance levels. Because the cones determine sensitivity at these levels, sensitivity is highest at the fovea and falls off monotonically with eccentricity (see Figure 14.7).

Color Perception. With large central defects eliminating substantial cone function, color perception is reduced. The observer must depend more on the rod system, which is monochromatic. However, significant difficulties present themselves when testing color vision in patients with macular disease because most of the tests, standards, and procedures have been developed for the CIE 2-degree standard observer (Knoblauch & Fischer, 1991). These tests tend to overestimate the degree of color vision loss in patients with macular disease (Knoblauch, Fischer, Robillard, & Faye, 1991).

Eye Movements. Eye movements and fixation stability appears to be affected by prechiasmatic central field losses primarily because of the new requirements they must fulfill as a result of the loss. For macular losses involving the fovea, for example, the observer must view eccentrically for most effective processing. For optimal function, saccades should be recalibrated to image objects of regard on some portion of the healthy nonfoveal retina. Patients must learn to secure a stable eccentric gaze

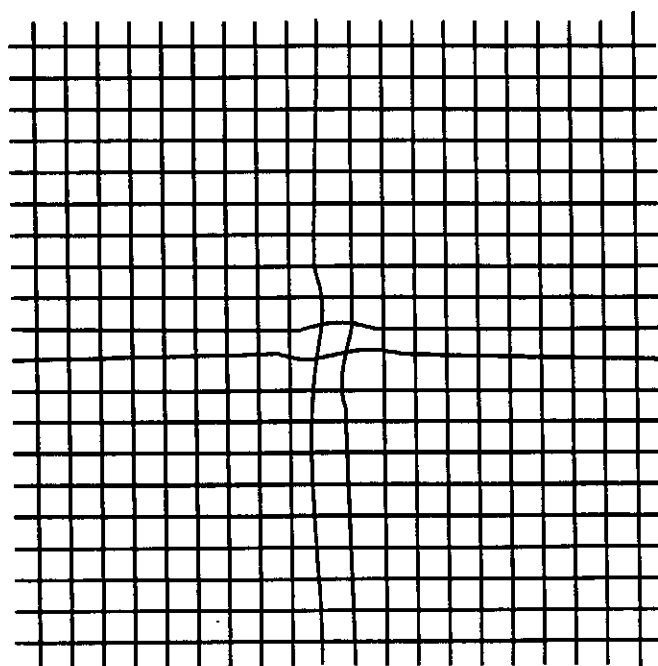


Figure 14.8. A regular grid may appear distorted to the person with foveal field defects. Such distortions (metamorphopsia) are also associated with errors of location, such as reduced bisection and vernier acuities.

while maintaining the object of focal attention on this area of retina and to pursue moving objects with a constant deviation of eye position relative to that required by a healthy macula. To what extent this is possible is not clear. However, most patients with foveal loss in macular disease do seem to develop preferred retinal loci for reading (Schuchard & Fletcher, 1994). However, islands of vision may be sufficient in size to carry out some tasks with one particular eccentric gaze, while other tasks may require a more eccentric eye position. Similarly, some areas of depressed sensitivity may be sufficient for performing visual tasks with high-contrast stimuli, whereas lower-contrast stimuli require using a wholly different portion of the retina. The fact that vergence eye movements may change the location and extent of volume scotomas further complicates this picture (Arditi, 1988).

Peripheral Retinal Field Defects

Peripheral retinal field defects fall outside the central 30 degrees. Generally speaking, these do not affect visual tasks requiring good spatial resolution or object localization. When they affect one eye alone, they may have little measurable effect on visual performance and little noticeable effect for the individual. However, bilateral peripheral field defects may decrease the likelihood of detecting obstacles during mobility, especially if they are located in the inferior visual field, where most potential obstacles are located. Chapter 15 treats difficulties with mobility associated with peripheral field loss.

The most common disorders of middle and later life producing peripheral retinal defects are glaucoma and retinitis pigmentosa (RP). Glaucoma usually begins with field losses outside the macula but within the central 30 degrees. It then progresses to arcuate central scotomas and, in later stages, to extensive peripheral and central losses. The early stages often go undetected for years, even when significant visual field losses occur. This fact shows that individuals with modest peripheral and midcentral losses do not experience specific functional difficulties associated with their losses. Extended zones of depressed or zero-contrast sensitivity will reduce the probability of detecting targets imaged in those zones, and visual performance will be affected even if not noticed by the patient.

RP is a disease with several subtypes that, rather than affecting local regions of the retina, differentially affects the photoreceptor types. Because it affects photoreceptor types rather than a visual field area, the disease does not easily fit into the classification based on visual field location, as described previously. The most common of the disease subtypes first attacks the rods and reduces visual function at night and under conditions of low luminance. Since the rod system is affected first, field losses tend to begin as a ring scotoma where the rods are most numerous—in the midperiphery (see Figure 14.7). They progress centrally and peripherally until all, or nearly all, vision is lost. Twelve distinct types of RP have been identified. Visual field loss can begin at any age (but most typically between ages nine and forty-five). It progresses with exponential decay of the visual field area. The time constant of the decay (4.3 years) appears to be the same for all patients (Massof & Finkelstein, 1987). Difficulties with night vision and with dark adaptation are the most prominent early symptoms of RP. Scotomas of moderate size that result from RP cause few functional difficulties until they encroach on the central 10 degrees, at which time the patient may experience difficulty with mobility.

Postchiasmatic Neural Deficits

Brain injury can cause different visual disorders ranging from visual field defects to impairments in cognitive visual abilities (for example, visual recognition and reading). Experimental and clinical evidence strongly suggests that different aspects of visual perception may be subserved by separate areas in the visual cortex (see Zeki, 1993, for an excellent review of this topic). Depending on the main location of acquired focal brain injury, patients may show particular visual disorders (for example, impairment or loss of color or movement vision). To simplify matters, occipitoparietal visual areas (the dorsal route) are thought to subserve visual space perception and cognition, while occipitotemporal visual areas (the ventral route) are assumed to subserve the processing of features, objects, faces, and scenes. The concept of functional specialization of the visual cortex does not imply, however, that every visual function has its own visual cortical area and that one visual cortical area subserves only one visual function. Indeed, selective vision losses oc-

cur rarely. On the other hand, brain injury is usually not restricted to only one visual cortical area. As a consequence, clusters of visual deficits (as distinguished from single deficits) are expected in the majority of patients with posterior brain injury.

Cerebral Vision Impairment

Unilateral Visual Field Disorders

Injury to the postchiasmatic visual pathway causes loss of vision in both monocular hemifields contralateral to the side of brain injury. The resulting homonymous visual field defects are typically incongruous in cases with injury to the anterior part of the postchiasmatic pathway. Congruous homonymous field loss, however, most frequently occurs with lesions affecting the posterior part of the optic radiation and the striate cortex (Harrington, 1976), presumably because nerve bundles serving corresponding positions of the visual fields are in poorer spatial registration in the anterior portion of the radiation. After unilateral brain injury, vision is lost in the hemifield (hemianopia), the upper or lower quadrant (quadrantanopia), or the parafoveal region (paracentral scotoma) (Figure 14.9). Unilateral homonymous hemianopia is by far the most

frequent type of homonymous field loss, followed by quadrantanopia and paracentral scotoma. Furthermore, visual field sparing is less than 5 degrees in the majority of patients; these patients typically have difficulties with reading and visual orientation. Tables 14.1 and 14.2 give a synopsis of the frequency of occurrence of the various unilateral and bilateral visual field defects and the degree of field sparing. The degree of insight among patients with homonymous field loss may vary between full insight and absence of any awareness (Critchley, 1949).

Spontaneous recovery from unilateral homonymous field loss can be expected in about 15% of cases within the first three months. The extent of recovery may range from a few degrees to complete recovery. The latter seems to occur only in exceptional cases (Zihl & Kennard, 1996).

Apart from absolute loss of vision in a particular part of the visual field, postchiasmatic brain injury may also cause a relative loss of visual functions in the affected field region. This type of homonymous field disorder is called *hemiambyopia*, a term first introduced by Poppelreuter (1917–1990). Light sensitivity may be only slightly depressed in some patients but severely depressed in others; color and form vision is always lost in the affected field

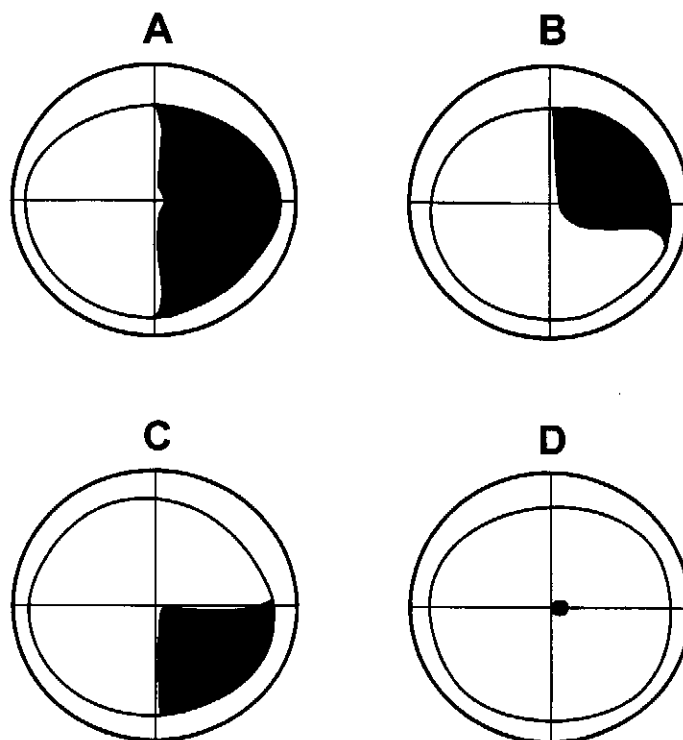


Figure 14.9. Forms of homonymous visual field loss in patients with damage to the central visual system. Black areas indicate binocular field loss. (A) Right-sided hemianopia. (B) Right upper quadrantanopia. (C) Right lower quadrantanopia. (D) Right-sided paracentral scotoma.

Table 14.1. Type and Frequency of Visual Field Disorders ($n = 568$)

Unilateral: 513 (90.3%); Bilateral: 55 (9.7%)

	Unilateral (%)	Bilateral (%)
Hemianopia	67.6	48.4
Upper quadrantanopia	8.2	5.4
Lower quadrantanopia	6.6	5.4
Paracentral scotoma	7.0	16.0
Hemiambyopia	10.6	10.6
Central scotoma	0.0	14.2

Source: Modified from Zihl, 1994.

regions (Figure 14.10). Moving and flickering targets can usually be detected and located without difficulty, provided their size and contrast are sufficiently high. Patients may also show a combination of visual field loss and amblyopic field region. In a group of 517 patients with unilateral posterior brain injury, Zihl (1994) found 11% of cases with homonymous hemiambyopia. Only one-quarter of the patients showed at least 5 degrees of field sparing; these patients exhibited reading difficulties similar to those in patients with parafoveal field loss. Hemianopic loss of color vision is discussed later in this chapter.

Bilateral Visual Field Loss

After bilateral postchiasmatic injury, vision may be lost in the left and right or upper and lower hemifields, or patients may show bilateral paracentral scotomas (Figure 14.11). Central vision is preserved in these patients, but it may be restricted to only a few degrees in diameter. As a consequence, patients may experience great difficulties with visual orientation, visual exploration, and reading. Since in many of these cases foveal vision is also impaired, their visual handicap is, as a rule, very severe. Vision can also be lost in the central field region while

Table 14.2. Visual Field Sparing in Patients with Unilateral Homonymous Field Disorders (in percent)

	< 2°	2°–4°	5°–10°	> 10°
Hemianopia	32.7	42.2	17.9	7.2
Quadrantanopia	17.1	42.1	29.0	11.8
Paracentral	40.5	40.5	19.0	0.0
Hemiambyopia	36.8	38.6	24.6	0.0

Source: Modified from Zihl, 1994.

being preserved in the periphery. These patients usually have great difficulties with visual orientation. Visual identification, recognition, and reading are severely impaired.

Complete Cerebral Blindness

In the case of complete destruction of optic radiations and/or striate cortices, vision is completely lost in the entire field. These patients may not always be aware of their blindness and may even deny it, claiming that their vision is normal (Anton's syndrome or anosognosia for blindness). About two-thirds of patients show recovery; the rest remain permanently blind (Zihl & Kennard, 1996). The time course and the extent of recovery can vary considerably. In a group of eighty-one cases, complete recovery of vision was observed in only four (6%). In the rest of the patients, partial recovery occurred within the first three months. It ranged from a return of light perception and a crude form vision to recovery of part of the visual field with or without form and color vision (Symonds & MacKenzie, 1957).

Visual Acuity and Spatial Contrast Sensitivity

After unilateral brain injury, visual acuity does not seem to be impaired except in cases where the optic tract is involved. In these patients, visual acuity may be reduced either in the eye ipsilateral to the site of brain injury or in both eyes (Savino, Paris, Schatz, & Corbett, 1978). However, as Frisén (1980) has argued, a relative acuity of even 90%, evaluated as being within the normal range, may in fact already represent a reduction. This would occur because the (usually unknown) relative acuity of the patient prior to brain injury might have been 100% or higher. Thus, relative acuity levels below 100% that cannot be improved by optical correction may be the result of unilateral postchiasmatic brain injury. Patients with bilateral brain injury may show either normal or nearly normal visual acuity. However, they can also exhibit a marked reduction in acuity, leaving them with only a crude form vision (Symonds & McKenzie, 1957).

Patients with unilateral or bilateral postchiasmatic brain injury sometimes complain of blurred or foggy vision, although visual acuity need not be im-

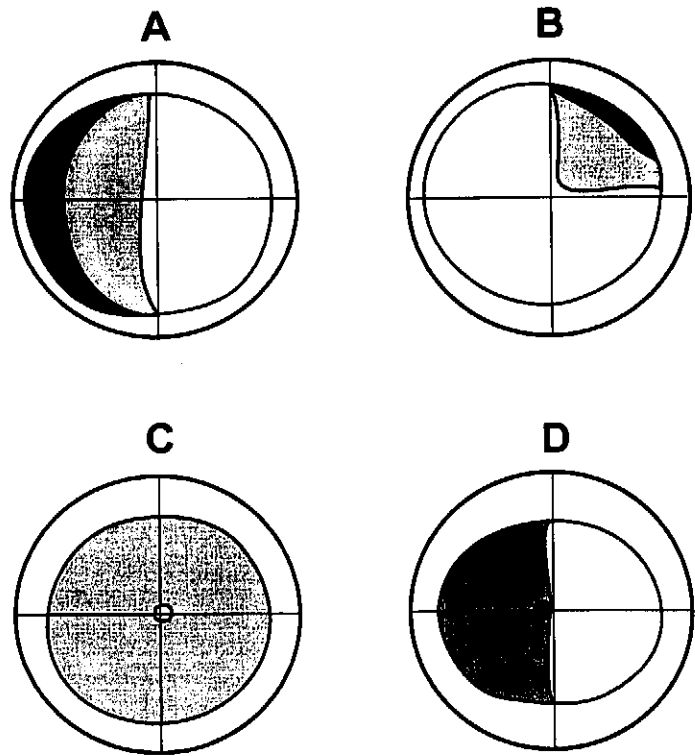


Figure 14.10. Forms of cerebral hemiamblyopia. Amblyopic regions are indicated in gray; lost field regions are shown in black. (A) Left-sided hemiamblyopia associated with loss of the temporal crescent of the left monocular field. (B) Amblyopia in the right upper quadrant. (C) Bilateral amblyopia with sparing of the central visual field. (D) Left-sided hemiamblyopia associated with left paracentral scotoma.

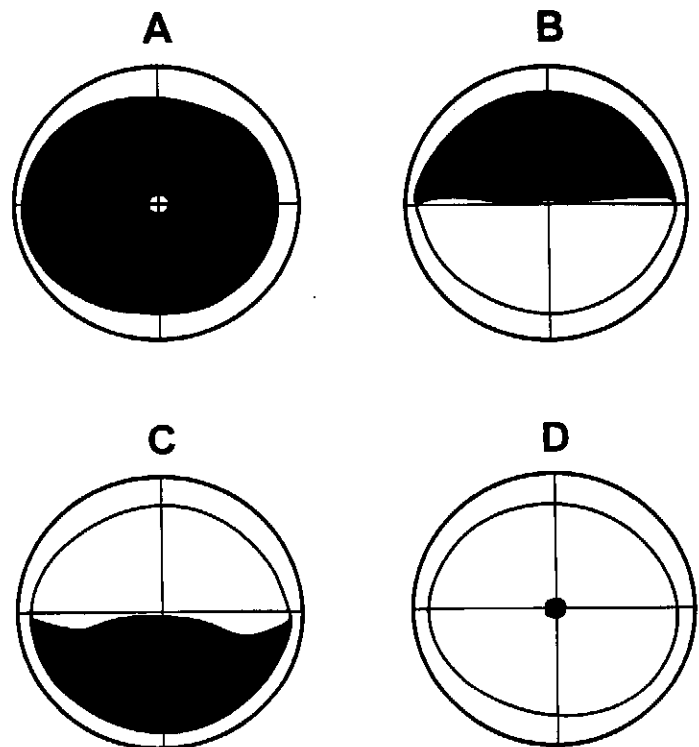


Figure 14.11. Forms of bilateral visual field loss after brain damage. Black areas indicate binocular field loss. (A) Bilateral homonymous hemianopia (tunnel vision). (B) Bilateral upper or superior quadrantanopia (altitudinal hemianopia). (C) Bilateral lower or inferior hemianopia. (D) Central scotoma.

paired and accommodation and convergence are not affected. These patients report reading difficulties; the letters appear to be indistinct and merge into one another. They also may find recognizing faces difficult, especially in black-and-white photographs, because contours appear very pale. Bodis-Wollner and Diamond (1976) showed that spatial contrast sensitivity is significantly reduced in such patients (Figure 14.12). Hess, Zihl, Pointer, and Schmid (1990) and Bulens, Meerwaldt, van der Wildt, and Keemink (1989) reported similar observations in larger groups of patients with posterior brain injury.

In some patients, vision is normal and clear on initial testing but deteriorates shortly afterward. This phenomenon has been interpreted in terms of fluctuation of vision due to increased fatigue (Bender & Teuber, 1946). This fluctuation can be assessed by recording pattern-generated, visually evoked potentials during a longer period of stimulation.

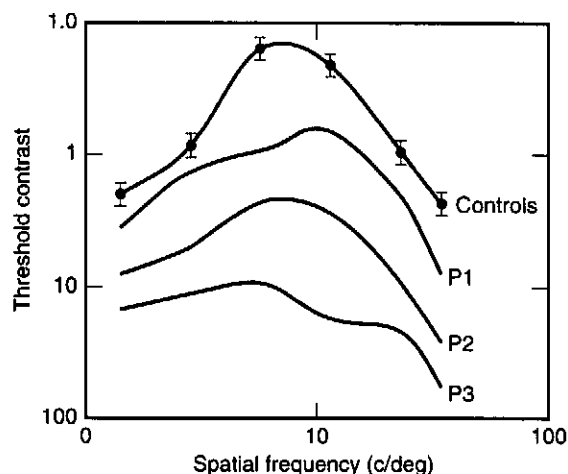


Figure 14.12. Binocular spatial contrast sensitivity (expressed as threshold contrast, mesopic conditions) in three male patients with unilateral occipital infarction. *P1* (fifty-six years old; duration of brain damage: eleven weeks) did not complain of visual blurring despite reduced contrast sensitivity. *P2* (fifty-two years old; duration of brain damage: thirteen weeks) complained of moderate visual blurring. *P3* (fifty-four years old; duration of brain damage: eleven weeks) reported severe visual blurring. For comparison, the results of a group of twenty control subjects aged between forty and sixty years are shown. Visual acuity for optotypes was 1.0 in *P1* and *P2* and only slightly reduced (0.9) in *P3*. (Data from Bodis-Wollner and Diamond, 1976)

Patients with delayed visual blurring may show increased fluctuation of the amplitude of the first positive wave of visually evoked potentials (Zihl & Schmid, 1989) (Figure 14.13).

Light and Dark Adaptation

Impairments in light and dark adaptation typically occur in patients with injury to the peripheral visual system. However, evidence shows that acquired brain injury can also impair light and/or dark adaptation. Patients with impaired light adaptation complain of being blinded under normal and sometimes even low illumination. They find it disturbing to look at, for example, a white sheet of paper because its reflection of light causes immediate blinding, and written words appear blurred or faded. Patients with impaired dark adaptation usually report that the surroundings appear dark. As a consequence, objects, buildings, or people cannot be seen clearly. Printed text and photographs appear grayish. Patients may claim that they need more light for reading and for recognizing their surroundings. In a study with 116 patients, Zihl and Kerkhoff (1990) found impairment of light and/or dark adaptation in the majority (78%) of cases, with the resulting disability being particularly severe. On the one hand, these patients need higher

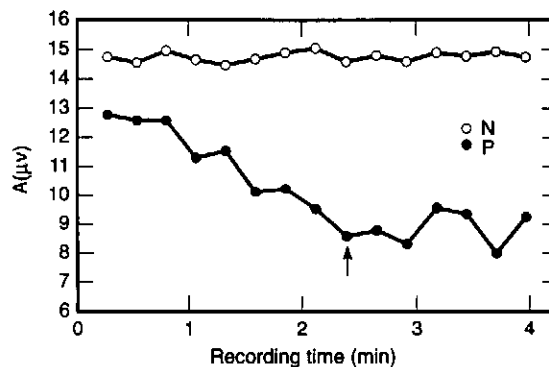


Figure 14.13. Course of amplitudes of the first positive wave of continuously recorded, pattern reversal, visually evoked potential (VEP) responses under binocular viewing conditions in a patient ($P = \bullet$) with closed head trauma (twenty-six years old, male; duration of brain damage: five weeks) and in a control subject ($N = \circ$; twenty-four years old, male). The arrow indicates the onset of visual blurring experienced by the patient (P). Note the decrease and fluctuation of the amplitude of the VEP in the patient.

illumination for clear vision. On the other, any small increase in illumination immediately produces a blinding sensation. Consequently, for reading, patients have problems identifying letters and words, either because the page is too dim and the letters do not appear to have sufficient contrast with their background, or because, at a higher luminance level, the entire (white) page looks as if it is "immersed in a glistening light." These phenomena occur for all types of visual stimuli in near vision, but patients are typically most aware of them when reading.

Color Vision

After brain injury, color vision can be affected in one hemifield (homonymous hemiachromatopsia) or may be impaired (dyschromatopsia) or even lost in the entire visual field, including the fovea (achromatopsia). Hemiachromatopsia occurs after unilateral posterior brain injury. It is defined as the loss of color vision in one hemifield, affecting either the entire hemifield or the upper quadrant. Patients are usually aware of this disorder and report that a part of the surroundings appears black and white or at least very pale. Mapping of the visual field with a light target does not reveal any field disorder. When a colored circle is moved from the periphery of the visual field toward its center, patients typically report a light target appearing in the periphery. They are able to identify its form at the respective eccentricity but not its color unless it crosses the border of the field where color vision is spared (Albert, Reches, & Silverberg, 1975; Damasio, Yamada, Damasio, Corbett, & McKee, 1980; Henderson, 1982). Note that form vision and light sensitivity are not impaired in the affected visual field region. This indicates that the loss of color vision is selective. Foveal color vision appears intact, but some patients may complain of a reduction of fine hue discrimination. Indeed, when tested with the Farnsworth-Munsell 100-hue test (Farnsworth, 1943), mild to moderate impairments in hue discrimination can be found (Zihl, Roth, Kerkhoff, & Heywood, 1988). Such impairments of color discrimination may not even be noticed by patients. However, they may represent a real visual disability if fine discrimination of colored hues is necessary for professional reasons.

In patients with bilateral brain injury, foveal color vision can be affected moderately (cerebral

dyschromatopsia) (Rizzo, Smith, Pokorny, & Damasio, 1993). In rare cases, it can even be lost (cerebral achromatopsia) (Meadows, 1974). Typically, all aspects where colors are involved are impaired: color discrimination, color constancy, color naming, and color association with objects (for example, yellow with banana). Patients with impaired or lost color vision in the entire field report that colored objects or pictures look pale, drained of color, or black and white. In contrast, the ability to differentiate grays correctly can be spared (Heywood, Wilson, & Cowey, 1987).

Visual Space Perception

Disorders of visual space perception comprise various types of impairment (for comprehensive reviews, see Benton & Tranel, 1993; De Renzi, 1982). Defective visual spatial localization is typically observed in the hemifield contralateral to the side of posterior brain injury, but it may also involve both hemifields. These patients exhibit difficulties in judging the position of an object in space relative to the observer (absolute position) or to other objects (relative position), which results in a defective localization of that object. Such patients may also have difficulties in properly shifting their gaze to a certain position in space and in accurately reaching for an object. Defective depth perception may affect depth localization in terms of underestimation or even overestimation of depth. This also affects size perception (micropsia in the case of underestimation; macropsia in the case of overestimation). Patients may even lose the experience of depth perception and stereopsis. A flight of stairs may then be perceived as a number of straight lines on the floor. Objects and faces may appear to have only two dimensions, especially in photographs. A further visuospatial impairment is the deviation of the subjective vertical, horizontal, and straight-ahead axes, which typically are shifted opposite to the site of brain injury (Figure 14.14A, B). The degree of deviation is larger after right-sided posterior brain injury, at least in right-handers. These patients find staying on the correct line difficult when writing, copying, and drawing. They also have difficulty maintaining the straight-ahead direction when walking or guiding their wheelchairs. Patients with visual neglect shift the straight-ahead direction toward the nonneglected side (Figure 14.14c). Finally,

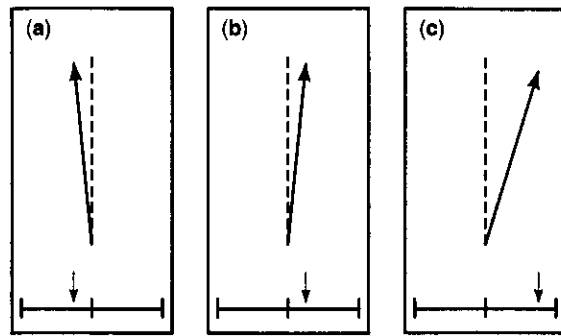


Figure 14.14. Shifts of straight-ahead direction in patients with left-sided (A; field sparing: 5 degrees) and right-sided (B; field sparing: 6 degrees) hemianopia and in a patient with left-sided visual neglect (C). At the bottom, the outcome of line bisection (length of line: 20 cm) is shown for each patient. The degree of shift of the straight-ahead direction was 4.3 degrees in A, 4.1 degrees in B, and 13.2 degrees in C. The shift in the line bisection task was 12 mm in A, 14 mm in B, and 17.6 mm in C. For comparison, normals bisected the line within a range of ± 5 mm.

patients with posterior brain injury may show difficulties with visual orientation (so-called visual disorientation). They complain of getting lost while reading or picture viewing. They find it difficult to gain and maintain an accurate orientation within visual stimulus arrays and visual scenes. As a consequence, they may also have problems with the spatial integration of visual impressions resulting from successive glances and with the spatial organization of visually guided oculomotor scanning.

Visual Cognition

Visual cognitive disorders are impairments in higher order or complex visual capacities, for example, visual recognition. Generally speaking, such capacities are based on visual-perceptual and cognitive functions and their interplay. With respect to diagnostics, it is important to find out whether an impairment or loss in visual recognition of an object or face can be explained by deficits in lower or elementary visual abilities or whether cognitive processes critically involved in visual recognition are affected. Impairments in the identification and/or recognition of visual material are summarized below. However, because considerable overlap exists between perception and recognition, a clear distinction between visual-perceptual and visual-cognitive

impairments is rarely possible. It therefore, appears reasonable and useful to describe higher-order vision impairments by specifying the underlying deficits. Indeed, careful and detailed testing and analysis of visual-perceptual capacities should always be part of the examination of a patient with visual-cognitive impairments. Such a diagnostic approach seems more appropriate since pure visual agnostic disorders occur relatively rarely.

In the following section, visual cognitive disorders are described with respect to the visual material that a patient can no longer recognize. Note that language and general intellectual functioning are preserved at sufficiently high levels, so they cannot account for the patient's failure to identify and recognize. When a patient fails to name visual material but can indicate visual recognition by other means—for example, by description or gesture—the failure is considered to be one of using verbal labels and is called *object anomia*.

Visual Object Agnosia

Patients with visual object agnosia have difficulties in identifying and recognizing objects visually but can do so in another modality (that is, when allowed to handle the object or to hear it in use). Typically, visual misidentification results from the incomplete or inappropriate use of object features. As a consequence, patients may confuse objects that share similar features. Often they use features not really characteristic for an object and do not consider other features. Thus, the cognitive deficits are (1) the impaired selection and integration of features that characterize an individual object and enable the differentiation from another similar object, and (2) the absence of processes that supervise visual identification and recognition, which could identify the faulty procedure (Damasio, Tranel, & Damasio, 1989; Grüsser & Landis, 1991).

Prosopagnosia

Prosopagnosia refers to the inability to recognize familiar faces visually and to learn new faces (Damasio et al., 1989; Grüsser & Landis, 1991). Most of the prosopagnosic patients reported in the literature also had difficulties with visually recognizing familiar objects (for example, animals, buildings, or cars). It is a striking condition. Patients can

no longer recognize the faces of their spouses, their children, or even themselves in a mirror or a photograph. They can usually, however, identify familiar people by their voices or even by their gait and other body movements. In a less severe condition, patients may exhibit difficulties differentiating between faces that look very similar and may confuse people easily.

Topographical Agnosia

The term *topographical agnosia* is often used to denote all difficulties with geographic orientation in the real world, in maps, or both. Patients suffering from this visual agnosia have problems orienting themselves in their familiar surroundings, and they cannot use maps or draw a plan of their house. Other patients may perform normally in their familiar surroundings, can drive long distances, and can navigate through city streets. However, they cannot show the corresponding routes on a map or locate the region or town in which they live.

Some patients complain of getting lost in familiar places, including their homes and neighborhood; this disturbance has been called *environmental agnosia*. In contrast with those experiencing topographical agnosia, these patients can correctly read maps and house plans, but they cannot make use of this ability for their spatial orientation. Topographical disorientation and environmental agnosia can occur separately but may also occur in association with each other (Grüsser & Landis, 1991).

Simultanagnosia

Patients with simultanagnosia have difficulty seeing more than one stimulus or more than a small portion of a scene or of a word at one time. The existence of this visual syndrome is still controversial, especially concerning its agnosic nature. Large visual field loss, visual neglect, and severe constriction of the field of attention impair or abolish the ability to get the complete overview, which is a crucial prerequisite for grasping the whole of an object, a face, a word, or a scene. Furthermore, the defective integration of several individual stimuli or features of an object into a whole may appear to be simultanagnosia but may, in fact, result from impaired visual feature integration due to visual object agnosia. Apparently, therefore, simultanagnosia can

be sufficiently explained as a visual-sensory, visual-attentional, or visual-cognitive impairment and need not be treated as a genuine visual agnosia (Grüsser & Landis, 1991).

Color Agnosia

This term has been coined to denote difficulties with the use of colors (except for naming). Despite having intact color discrimination, patients with color agnosia typically have problems associating colors with objects (for example, yellow with banana). They have difficulties coloring objects and do not reject absurd object coloring (for example, a red elephant). Color agnosia, like other visual agnosias, seems to occur rarely (Grüsser & Landis, 1991).

Dyslexia and Alexia

Reading can be impaired after brain injury at the visual-sensory, the visual-attentional, or the cognitive-semantic level. Difficulties at the visual-sensory level can interfere with reading because text information may be missing or disrupted. Since the parafoveal visual field region is essential for both the acquisition of words and the visual guidance of eye movements during reading, loss of vision in this portion of the visual field impairs reading in particular. Patients with left-sided visual field loss have difficulty shifting their gaze from right to left, impairing their ability to go from one end of a line to the beginning of the next. They may also omit prefixes. Patients with right-sided field loss typically omit suffixes and have problems guiding their eyes from left to right. Reading difficulties are related to the degree of visual field sparing: the smaller the degree of sparing, the more pronounced the impairment. Furthermore, patients with right-sided field loss are more impaired than patients with left-sided field loss. This difference is explained by the observation that readers of left-to-right orthographies acquire more information from the right side of fixation than from the left (Zihl, 1995b). Since homonymous hemianopia is the most frequent cause of parafoveal visual field loss, the resulting reading impairment is often called *hemianopic dyslexia*. In cases of visual hemineglect, reading is even more severely impaired. Patients with neglect dyslexia due to left-sided visual neglect typically omit all

the text on a page that lies to the left of their fixation and may also omit the prefixes of single words (Behrmann, 1994). In contrast with patients with hemianopic dyslexia, who benefit from being instructed to shift the gaze to the left or right, patients with visual neglect cannot make use of this information or do so only at a later stage of recovery. Interestingly, patients with hemianopic and with neglect dyslexias can read correctly when words are presented vertically in the unaffected hemifield.

Patients with pure alexia do not have difficulties at the visual-sensory or visual-attentional levels. However, they do have problems identifying individual letters and/or building words out of letters (letter-by-letter reading). The term *pure* refers to the fact that impaired reading in these patients is not part of a more general aphasic disturbance. The severity of impairment also depends on the character of the letters. Reading of handwritten words (even the patient's own) is typically more impaired than the reading of words in distinct print. In contrast with patients who have hemianopic and neglect dyslexia, those with pure alexia show no improvement in reading when words are presented vertically (Grüsser & Landis, 1991).

Visual Attention

Apart from general slowing, which is a common nonspecific sign after acquired brain injury and is characterized by an increased need for time in processing visual information (Cohen & O'Donnell, 1993), brain injury can cause specific disorders in visual attention. Unilateral visual inattention can be associated with homonymous visual field loss, typically hemianopia. It can affect either the left or the right hemifield. Patients with this type of visual inattention omit stimuli, especially in unfamiliar and complex situations. However, they have fewer difficulties in their familiar surroundings. Because of the loss of vision in one hemifield, these patients frequently have to use large gaze shifts to compensate for the field loss and to gain a quick and adequate overview. Patients with hemianopia exhibit particular difficulties when stimuli attract attention by appearing in that portion of the visual surrounding that corresponds to the preserved hemifield, thereby hindering the patient's ability to regard, at the same time, stimuli appearing in the affected hemifield. Some patients show a tendency to over-

compensate for the visual field loss and so may neglect stimuli in their good hemifield. Note that patients with hemi-inattention caused by hemianopic field loss and with problems in shifting attention to the affected side benefit from knowledge about the actual visual surroundings. They can then shift their attention and gaze toward it. Such patients have no, or only minor, difficulties in their own homes and can attend to the affected side when instructed to do so.

In contrast with patients having this type of visual hemi-inattention, patients with so-called visual neglect are not aware of their visual surroundings left or right of their line of gaze. Visual neglect typically affects the left side (at least in right-handed people). In its severest form, it is characterized by a rightward deviation of the eyes and the head. When asked to do so, patients fail to look to the left side and pick up objects lying left of their actual fixation position. When asked to copy an object (for example, a flower) or to draw it from memory, patients draw only the left half of the object. In a crossing-out test, patients neglect items mainly in the left portion of the page (Figure 14.15). Reading is also impaired because patients omit the left part of a line or a single word. When asked to write, patients use only the extreme right portion of a page. Patients are typically unaware of their visual neglect and may offer bizarre stories to explain their difficulties when confronted with the behavioral consequences (Grüsser & Landis, 1991; Heilman, Watson, & Valenstein, 1993).

At a later stage of recovery, patients may be able to shift their attention toward the affected side and to guide their eye and head movements to search for stimuli there. When a competing stimulus appears in the preserved hemifield, however, patients tend to shift their attention immediately to this new stimulus and to neglect, at the same time, stimuli appearing in the affected hemifield. When stimuli are presented simultaneously in both hemifields, the one appearing in the affected hemifield either undergoes extinction or the response is delayed (Heilman et al., 1993; Mattingley, Bradshaw, Bradshaw, & Nettleton, 1994).

Bilateral visual inattention is the most prominent symptom in patients with so-called Balint's syndrome (Grüsser & Landis, 1991). Stimuli outside the central visual field are not detected. Even if stimuli are in the field of vision, patients can see

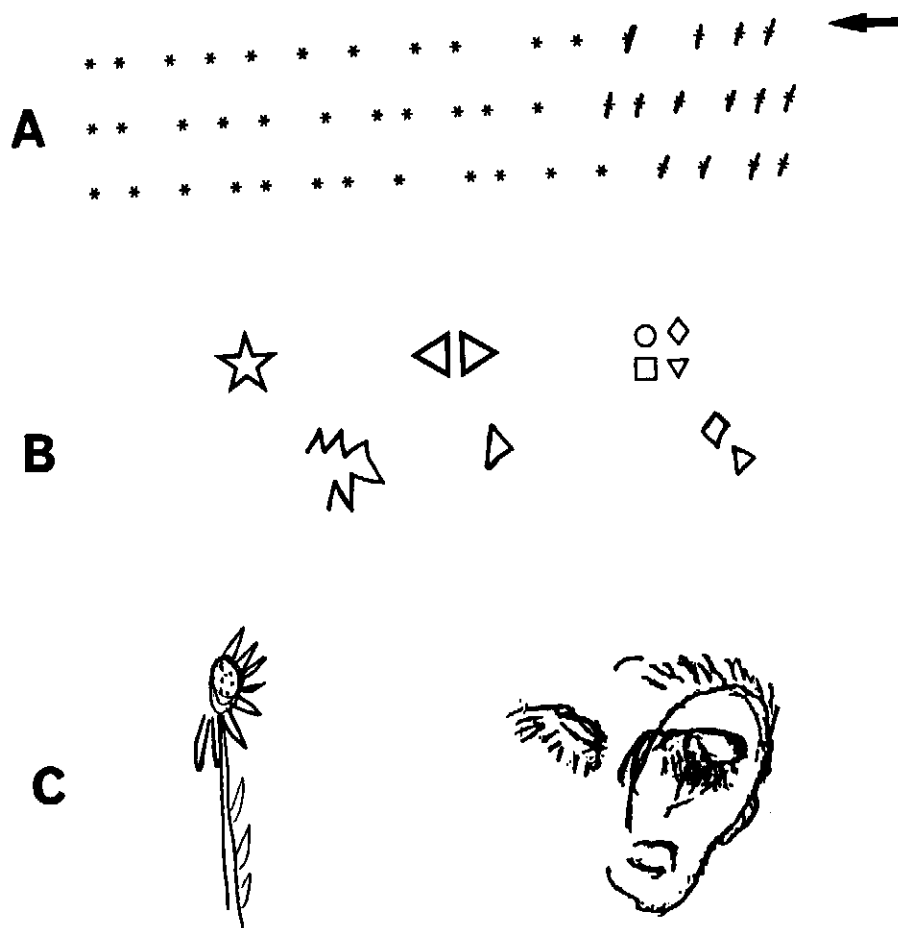


Figure 14.15. Cancellation (A); copying (B); and drawing from memory (C) in a patient with left-sided visual neglect. Note omissions of targets in A (the arrow indicates the direction of the patient's cancellation); omissions of the left half of figures and shift of drawings to the right in B; and omission of the left half of a flower and a face in C.

only one stimulus at a time. A stimulus that has entered the small field of vision seemingly catches the patient's attention, but he or she cannot shift the gaze to the stimulus even after detecting it. Patients are unable to shift their eyes voluntarily or on verbal command and show sticky fixation. In addition, they have striking problems with visual space perception—that is, they find estimating distances and directions in space to be difficult. As a consequence, visual guidance of ocular and hand motor responses when fixating objects or reaching and grasping for them is also affected. Thus, patients with Balint's syndrome experience bilateral visual inattention, impaired visual space perception, and the inability to initiate and make use of oculomotor and hand motor activities. Patients with this syndrome in its severest form are behaviorally blind. Patients with minor forms omit stimuli

in the entire hemifield, whereby the left-sided neglect is typically more pronounced. They also have difficulties perceiving the visual field as a whole.

Eye Movements

Brain injury can alter the accuracy of visually guided saccadic eye movements. It may also impair visually guided oculomotor visual scanning during the inspection of objects, faces, or scenes. Figures 14.16 and 14.17 show examples of patients with defective saccadic accuracy and impaired oculomotor scanning. Patients with homonymous visual field loss typically show hypometric saccades toward the affected side. In cases with additional visual disorientation, oculomotor scanning is often clumsy, remarkably slowed, and disorganized in both hemifields, even in patients with unilateral posterior brain in-

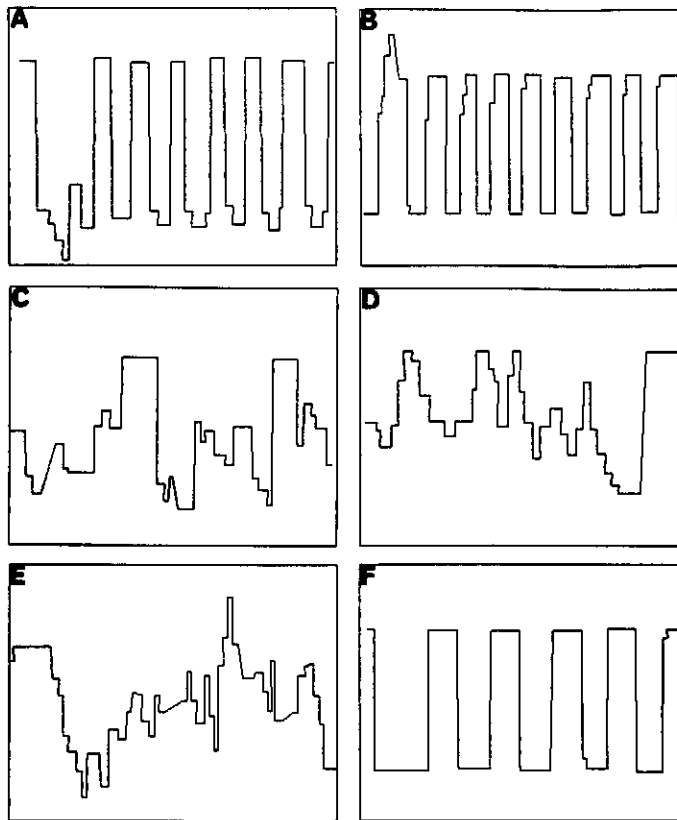


Figure 14.16. Voluntary horizontal eye movements to light targets separated by 20 degrees in patients with homonymous hemianopia (A: left-sided; B: right-sided; field sparing: 1 degree, in both cases); in a patient with bilateral hemianopia (C: diameter of spared central field: 4 degrees); in a patient with left-sided visual neglect (D); and in a patient with Balint's syndrome (E). Note hypometria of saccades to the affected side(s). Leftward and rightward eye movements are shown as higher and lower, respectively, on vertical axes. For comparison, voluntary horizontal saccades of a control subject are shown in F.

jury (Zihl, 1995a; Zihl & Hebel, 1997). In cases with left-sided visual neglect, hypometria is especially pronounced for saccades to the left. Oculomotor scanning activity is restricted to the right part of the surroundings and is characterized by spatial disorganization. When patients recover from visual neglect, they are able to shift their gaze to the affected side and can search for stimuli there. However, a residual right-sided bias can still be observed, even months later, which can elicit the symptoms of visual neglect (Karnath, 1988; Mattingley et al., 1994). Patients with bilateral visual inattention (Balint's syndrome) show highly inaccurate visually guided saccadic eye movements. Because they are unable to unlock fixation (spasmodic fixation), they have great difficulties in initiating saccades and shifting their gaze from one point in space to another. Saccadic gaze shifts may therefore be characterized by a pattern of erratic, sometimes wandering movements when searching for the target. Spontaneous saccades may be present or absent; on verbal command, saccades may be elicited in some patients but not in others (Pierrot-Deseilligny, Gray, & Brunet, 1986).

Summary

Neural visual disorders of middle and later life may be conveniently divided into those affecting the visual pathway before and after decussation of optic nerve fibers at the optic chiasm. This anatomic division also corresponds to the boundary between the clinical domains of the ophthalmologist and optometrist, on the one hand, and the neurologist and neuropsychologist on the other. In prechiasmatic disorders, functional effects are fairly well predicted by the visual field locations affected, despite a great variety of specific diseases and disorders that can affect the eye. Postchiasmatic disorders are uniform in the sense that they are all simple lesions, whether caused by traumatic brain injury, cerebrovascular accident, or tumor. However, in postchiasmatic disorders, predicting functional deficits from lesion localization within the brain is more difficult because higher-level visual functions depend on widely distributed processes. Postchiasmatic disorders are consequently better described by their associated functional deficits than by any anatomic description.

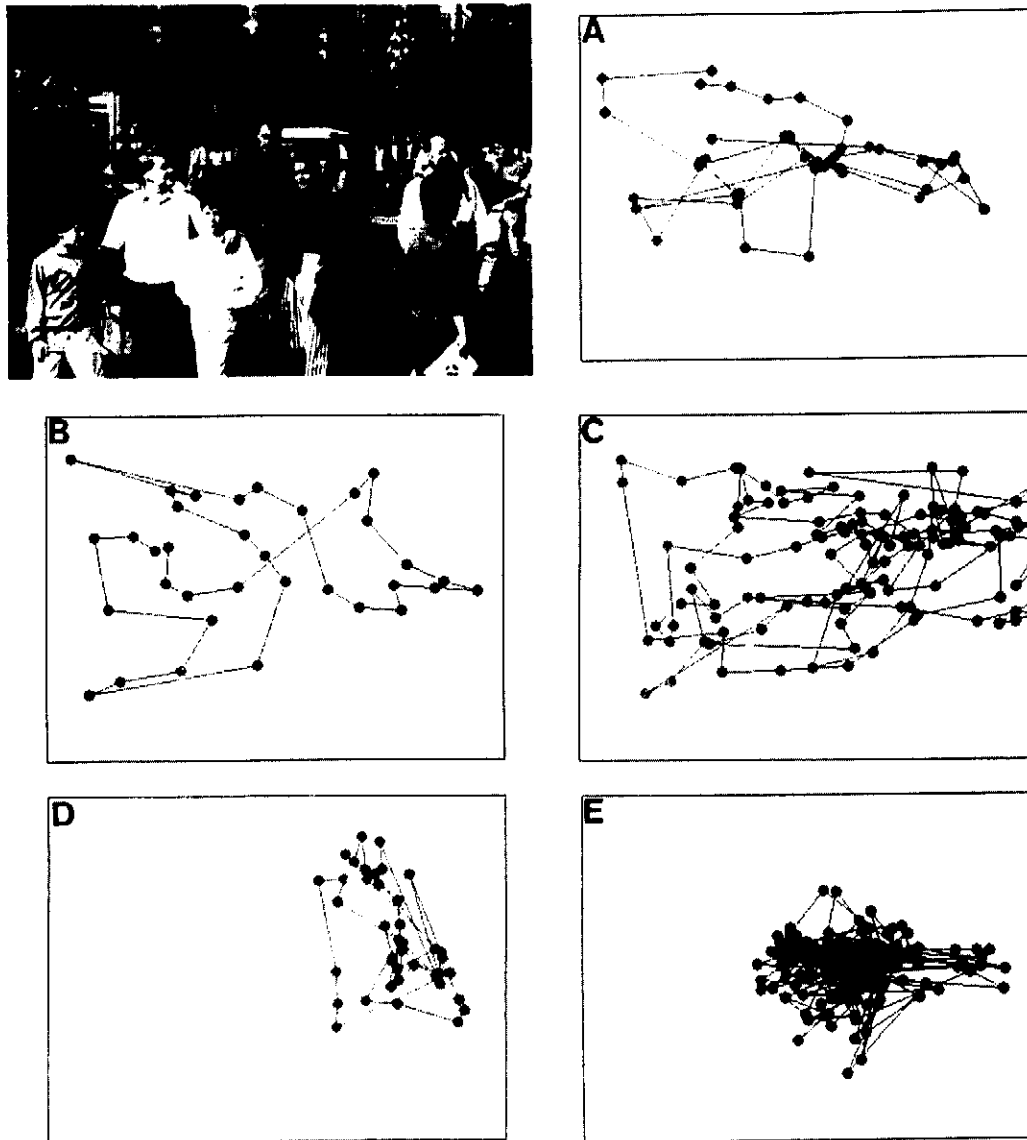


Figure 14.17. Oculomotor scanning patterns during the inspection of a scene (top left of the figure; size: 40 degrees horizontally, 30 degrees vertically) in brain-damaged patients and in a normal subject. Scanning times in parentheses. A: Control subject (17.1 sec.); B: patient with left-sided hemianopia (field sparing: 3 degrees) and very efficient oculomotor compensation (17.8 sec.); C: patient with visual disorientation (60.3 sec.); D: patient with left-sided visual neglect (56 sec.); and E: patient with Balint's syndrome (61 sec.). Note increased number of fixations in C and restricted fields of scanning in D and E.

Despite these differences, however, disruption of visual processing prechiasm and postchiasm may produce many of the same types of deficits, including in contrast sensitivity, light and dark adaptation, color perception, and space perception. And despite the clinical division between eye-care and neurological care, a common language is useful when considering all the neural deficits of vision.

REFERENCES

- Albert, M. L., Reches, A., & Silverberg, R. (1975). Hemianopic colour blindness. *Journal of Neurology, Neurosurgery, and Psychiatry*, 38, 546-549.
- Arditi, A. (1988). The volume visual field: A basis for functional perimetry. *Clinical Vision Sciences*, 3, 173-183.

- Baylor, D. A. (1987). Photoreceptors and vision. *Investigative Ophthalmology and Visual Science*, 28, 34-49.
- Behrmann, M. (1994). Neglect dyslexia: Attention and word recognition. In M. J. Farah & G. Ratcliff (Eds.), *The neuropsychology of high-level vision* (pp. 173-214). Hillsdale, NJ: Erlbaum.
- Bender, M. B., & Teuber, H. L. (1946). Phenomena of fluctuation, extinction and completion in visual perception. *Archives of Neurology and Psychiatry*, 55, 627-658.
- Benton, A., & Tranel, D. (1993). Visuo-perceptual, visuo-spatial, and visuo-constructive disorders. In K. M. Heilman & E. Valenstein (Eds.), *Clinical neuropsychology* (3rd ed., pp. 165-213). New York: Oxford University Press.
- Bodis-Wollner, I., & Diamond, S. P. (1976). The measurement of spatial contrast sensitivity in cases of blurred vision associated with cerebral lesions. *Brain*, 99, 695-710.
- Bulens, C., Meerwaldt, J. D., van der Wildt, G. J., & Keemink, D. (1989). Spatial contrast sensitivity in unilateral cerebral ischemic lesions involving the posterior visual pathway. *Brain*, 112, 507-520.
- Cohen, R. A., & O'Donnell, B. F. (1993). Disturbances of attention. In R. A. Cohen (Ed.), *The neuropsychology of attention* (pp. 255-274). New York: Plenum Press.
- Critchley, M. (1949). The problem of awareness or non-awareness of hemianopic field defects. *Transactions of the Ophthalmological Society UK*, 69, 95-109.
- Damasio, A. R., Tranel, D., & Damasio, H. (1989). Disorders of visual recognition. In F. Boller & J. Grafman (Eds.), *Handbook of neuropsychology* (Vol. 2, pp. 317-322). Amsterdam: Elsevier Science.
- Damasio, A., Yamada, T., Damasio, H., Corbett, J., & McKee, J. (1980). Central achromatopsia: Behavioral, anatomic, physiologic aspects. *Neurology*, 30, 1064-1071.
- DeRenzi, E. (1982). *Disorders of space exploration and cognition*. New York: Wiley.
- Esterman, B. (1982). Functional scoring of the binocular field. *Archives of Ophthalmology*, 89, 35-42.
- Farnsworth, D. (1943) The Farnsworth-Munsell 100-hue and dichotomous tests for color vision. *Journal of the Optical Society of America*, 33, 569-578.
- Frisén, L. (1980). The neurology of visual acuity. *Brain*, 103, 639-670.
- Grüsser, O. J., & Landis, T. (1991). Visual agnosias and other disturbances of visual perception and cognition. Boca Raton, FL: CRC Press.
- Harrington, D. O. (1976). *The visual fields* (4th ed.). St. Louis, MO: Mosby.
- Hecht, S., Schlaer, S., & Pirenne, M. H. (1942). Energy, quanta and vision. *Journal of General Physiology*, 25, 819-840.
- Heilman, K. M., Watson, R. T., & Valenstein, E. (1993). Neglect and related disorders. In K. M. Heilman & E. Valenstein (Eds.), *Clinical neuropsychology* (3rd ed., pp. 279-336). New York: Oxford University Press.
- Henderson, V. W. (1982). Impaired hue discrimination in homonymous visual fields. *Archives of Neurology*, 39, 418-419.
- Hess, R. F., Zihl, J., Pointer, J. S., & Schmid, C. H. (1990). The contrast sensitivity deficit in cases with cerebral lesions. *Clinical Vision Sciences*, 5, 203-215.
- Heywood, C. A., Wilson, B., & Cowey, A. (1987). A case study of cortical color "blindness" with relatively intact achromatic discrimination. *Journal of Neurology, Neurosurgery, and Psychiatry*, 50, 22-29.
- Hood, D. C., & Finkelstein, M. A. (1986). Sensitivity to light. In K. Boff, L. Kaufman, & J. Thomas (Eds.), *Handbook of perception and human performance: Vol. 1. Sensory processes and perception*. New York: Wiley.
- Karnath, H. O. (1988). Deficits in attention in acute and recovered visual hemi-neglect. *Neuropsychologia*, 26, 27-43.
- Knoblauch, K., & Fischer, M. (1991). Low vision issues in color vision. In B. P. Rosenthal & R. G. Cole (Eds.), *A structured approach to low vision care* (Vol. 3). Philadelphia: Lippincott.
- Knoblauch, K., Fischer, M., Robillard, N., & Faye, E. (1991). The effect of test element size on performance of the D-15 in age-related maculopathy and congenital color deficiency. In B. Drum, J. D. Moreland, & A. Serra (Eds.), *Colour vision deficiencies IX* (Vol. 54). Dordrecht, Netherlands: Kluwer.
- Kosnik, W., Fikre, J., & Sekuler, R. (1986). Visual fixation stability in older adults. *Investigative Ophthalmology and Visual Science*, 27, 1720-1725.
- Levi, D. (1991). Spatial vision in amblyopia. In D. Regan (Ed.), *Spatial vision*. Boca Raton, FL: CRC Press.
- Massof, R. W., & Finkelstein, D. (1987). A two-stage hypothesis for the natural course of retinitis pigmentosa. In E. Zrenner, H. Krastel, & H. H. Goebel (Eds.), *Research in retinitis pigmentosa*. Oxford: Pergamon Press.
- Mattingley, J. B., Bradshaw, J. L., Bradshaw, J. A., & Nettleton, N. C. (1994). Residual rightward attentional bias after apparent recovery from right hemisphere damage: Implications for a multicomponent model of neglect. *Journal of Neurology, Neurosurgery, and Psychiatry*, 57, 597-604.
- Meadows, J. C. (1974). Disturbed perception of colors associated with localized cerebral lesions. *Brain*, 97, 615-632.
- Pierrot-Deseilligny, C. H., Gray, F., & Brunet, P. (1986). Infarcts of both inferior parietal lobules with impairment of visually guided eye movements,

- peripheral visual inattention and optic apraxia. *Brain*, 109, 81-97.
- Poppelreuter, W. (1990). *Disturbances of lower and higher visual capacities caused by occipital damage* (J. Zihl & L. Weiskrantz, Trans.). Oxford: Clarendon Press. (Original work published 1917)
- Rizzo, M., Smith, V., Pokorny, J., & Damasio, A. R. (1993). Color perception profiles in central achromatopsia. *Neurology*, 43, 995-1001.
- Savino, P. J., Paris, M., Schatz, N. J., & Corbett, J. J. (1978). Optic tract syndrome. *Archives of Ophthalmology*, 96, 656-663.
- Schuchard, R. (1991). Perception of straight lines across a scotoma. *Investigative Ophthalmology and Visual Science*, 32S, 816.
- Schuchard, R. A., & Fletcher, D. C. (1994). Preferred retinal locus: A review with applications in low vision rehabilitation. *Ophthalmology Clinics of North America*, 7, 243-256.
- Sunness, J. S., Bressler, N. M., & Maguire, M. G. (1995). Scanning laser ophthalmoscopic analysis of the pattern of visual loss in age-related geographic atrophy of the macula. *American Journal of Ophthalmology*, 119, 143-151.
- Symonds, C., & MacKenzie, I. (1957). Bilateral loss of vision from cerebral infarction. *Brain*, 80, 415-455.
- Westheimer, G. (1979). The spatial sense of the eye (Proctor Lecture). *Investigative Ophthalmology and Visual Science*, 18, 893-912.
- Zeki, S. (1993). *A vision of the brain*. Oxford: Blackwell Scientific.
- Zihl, J. (1994). Rehabilitation of visual impairments in patients with brain damage. In A. C. Kojman et al. (Eds.), *Low vision: Research and new developments in rehabilitation* (pp. 287-295). Amsterdam: IOS Press.
- Zihl, J. (1995a). Eye movement patterns in hemianopic dyslexia. *Brain*, 118, 891-912.
- Zihl, J. (1995b). Visual scanning behavior in patients with homonymous hemianopia. *Neuropsychologia*, 33, 287-303.
- Zihl, J., & Hebel, N. (1997). Patterns of oculomotor scanning in patients with unilateral posterior parietal or frontal lobe damage. *Neuropsychology*, 35, 893-906.
- Zihl, J., & Kennard, C. (1996). Disorders of higher visual function. In T. Brandt et al. (Eds.), *Therapy and course of neurological disorders* (pp. 201-212). New York: Academic Press.
- Zihl, J., & Kerkhoff, G. (1990). Foveal photopic and scotopic adaptation in patients with brain damage. *Clinical Vision Sciences*, 5, 185-195.
- Zihl, J., Roth, W., Kerkhoff, G., & Heywood, C. A. (1988). The influence of homonymous visual field disorders on color sorting performance in the FM 100-hue test. *Neuropsychologia*, 26, 869-876.
- Zihl, J., & Schmid, C. (1989). Use of visually evoked responses in evaluation of visual blurring in brain-damaged patients. *Electroencephalography and Clinical Neurophysiology*, 74, 394-398.