P. M., a colonel in the British army who fought in North Africa during the Second World War, was struck by a bullet that went through the back of his brain. Miraculously, P. M. survived, but his vision was severely affected. He completely lost sight in the right visual field but the central part of his left visual field survived. He reported that he could see "normally" in a region of the left visual world that was about the diameter of your fist held at arm's length directly in front of your face.

P. M.'s symptoms reveal a topographical map of the visual world in the occipital cortex and the possibility of seeing through only a small part of it. But what did P. M. experience in the rest of the visual world? Shortly after his injury, he reported that the lost world appeared black, as though the lights were out. Occasionally, however, he was aware that the lost regions were different, "almost gray," although he could never express specifically what exactly was different other than the grayness.

P. M. also experienced a phenomenon that many patients with extensive visual-field defects experience: if asked to guess whether a spot of light had blinked in his blind field, he could "guess" at above-chance levels. He was not consciously aware that the light had appeared and seemed bemused that he could guess, sometimes quite accurately, about the presence or absence of the light.

In spite of his residual central vision, P. M. had two particular (and, for him, aggravating) problems: he found it very difficult to read and he had difficulty recognizing faces. Curiously, however, P. M. could recognize other objects more easily, even though he could not see any more of them than he could the faces.

Cases such as P. M.'s are especially interesting because our brains are organized around vision. Our perception of the world is predominantly visual, our movements are guided by visual information, our social and sexual behavior is highly visual, our entertainment is largely visual, and our nights

are enriched by visual dreams. In this chapter, we first consider the anatomical organization of the occipital lobes and then examine the extent of the visual system within the brain. Next, we examine disorders of the visual pathways and of the visual system. We shall see why faces present a special problem for the visual system and why the ability to visualize presents humans a unique opportunity.

Anatomy of the Occipital Lobes

The occipital lobes form the posterior pole of the cerebral hemispheres, lying under the occipital bone at the back of the skull. On the medial surface of the hemisphere, the occipital lobe is distinguished from the parietal lobe by the parieto-occipital sulcus, as illustrated in Figure 13.1. No clear landmarks separate the occipital cortex from the temporal or parietal cortex on the lateral surface of the hemisphere, however, because the occipital tissue merges with the other regions. The lack of clear landmarks makes it difficult to define the extent of the occipital areas precisely and has led to much confusion about the exact boundaries.

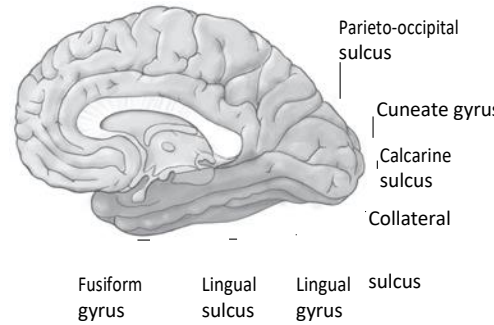


Figure 13.1 Medial view of the occipital lobe, illustrating the major landmarks.

Within the visual cortex, however, are three clear landmarks. The most prominent is the calcarine sulcus, which contains much of the primary cortex (see Figure 13.1). The calcarine fissure divides the upper and lower halves of the visual world. On the ventral surface of the hemisphere are two gyri (lingual and fusiform). The lingual gyrus includes part of visual cortical regions V2 and VP, whereas V4 is in the fusiform gyrus.

Subdivisions of the Occipital Cortex

The monkey cortex was first divided by Brodmann into three regions (areas 17, 18, and 19, see Figure 10.7), but studies using imaging, physiological, and newer anatomical techniques have produced much finer subdivisions. Although the map is still not complete, the consensus is that the occipital cortex contains at least nine different visual areas, as illustrated in Figure 13.2: V1, V2, V3, VP, V3a, V4d, V4v, DP, and MT (also known as V5).

Figure 13.2 shows the locations of some of these areas on the lateral surface of the monkey brain as well as their locations on a two-dimensional flattened map that includes both the lateral areas and those

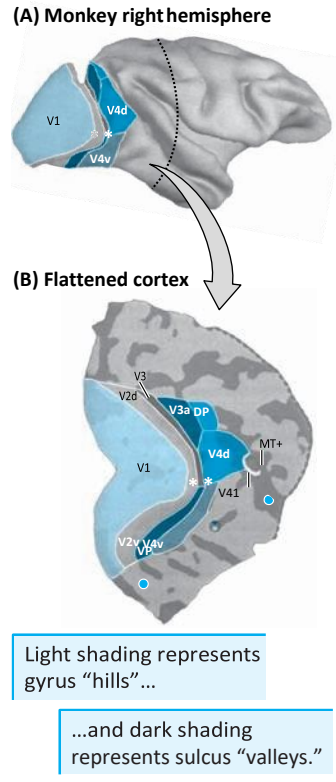


Figure 13.2 Topography of the visual cortex of the macaque monkey. (A) A nearly normal rendition of the lateral surface of the brain in which the sulci are opened slightly. (B) A flattened cortical surface showing both the lateral and the medial regions. The shaded areas represent regions that are normally curved up (gyri) or down (sulci). The asterisks refer to the foveal representation in areas B1 and V4. (After Tootell and Hadjikhani, 2001, with permission.)

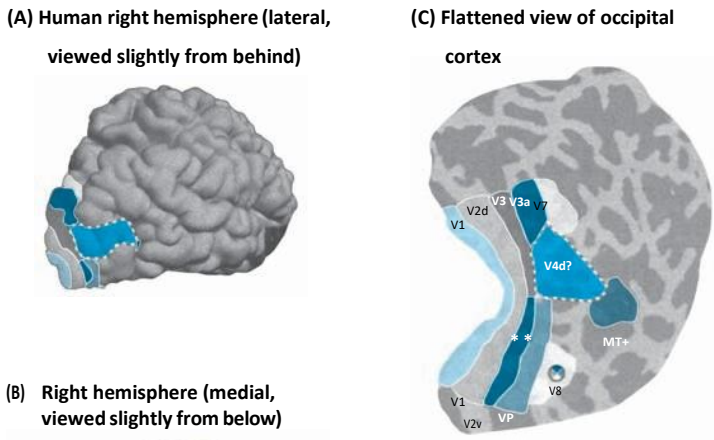


Figure 13.3 Topography of the human visual cortex. (A) A nearly normal rendition of the lateral surface of the brain in which the sulci are opened slightly. (B) The medial surface in which the sulci are opened slightly. (C) A flattened cortical surface showing both the lateral and medial regions. The shaded areas represent regions that are normally curved up (gyri) or down (sulci). The asterisks refer to the foveal representation in areas V1 and V4. (After Tootell and Hadjikhani, 2001, with permission.)

located on the medial surface of the hemisphere. The precise locations of the human homologues are still not settled, but Figure 13.3 presents a recent map constructed by Tootell and Hadjikhani that includes all of the monkey areas as well as an additional color-sensitive area (V8).

Some of the areas contain a complete visual field, whereas others have only an upper or lower visual field. This distinction is curious, and Previc has suggested that the upper and lower fields may have different functions, with the upper more specialized for visual search and recognition and the lower more specialized for visuomotor guidance.

A remarkable feature of area V1 is its complex laminar organization, which, as illustrated in Figure 13.4, is probably the most distinct of all cortical areas. Although we usually say that the cortex has six layers, it is possible to see many more in area V1, partly because layer IV alone features four distinct layers. Another surprising feature of area V1 is that, although it appears to be anatomically homogenous, it can be shown to actually be heterogeneous by staining it for the enzyme cytochrome oxidase, which is crucial in making energy available to cells. Regions of cytochrome-rich areas, known as blobs, are separated by interblob regions of little cytochrome activity (see Figure 10.10). It turns out that cells in the blobs take part in color perception and the interblobs have a role in form and motion perception.

The discovery that area V1 is functionally heterogeneous—that a given cortical area may have more than one

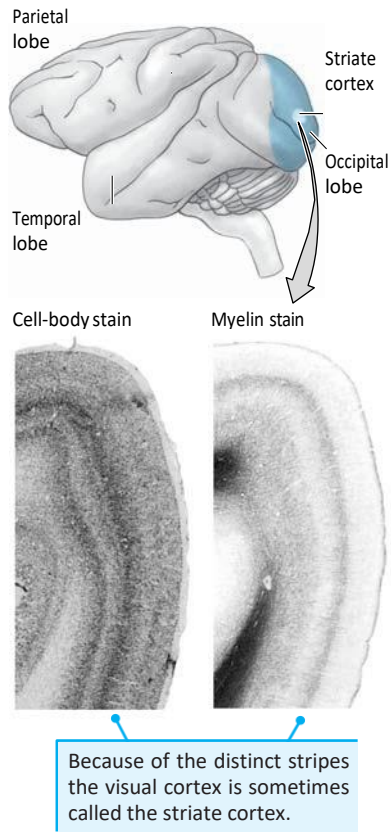


Figure 13.4 The visual cortex is highly laminated, as can be seen in a cell body stain (left) or a myelin stain (right) in these sections from a monkey brain. Because of the distinct stripes, the visual cortex is sometimes called the striate cortex.

distinct function—
was unexpected. It

turns out that area V2 also is heterogeneous when stained with cytochrome oxidase, but, instead of blobs, stripes are revealed (see Figure 10.10). One type, the "thin stripe," takes part in color perception. Two other types, known as thick stripes and pale stripes, have roles in form and motion perception, respectively. Thus, we see that the heterogeneity of function observed in area V1—representing color, form, and motion—is preserved in area V2, although it is organized in a different way.

The distribution of color function across much of the occipital cortex and beyond (i.e., areas V1, V2, V4, V8) is important because, until recently, the perception of form or movement was believed to be colorblind. It has now become clear that color vision is integral to the analysis of position, depth, motion, and structure of objects (see a review by Tanaka et al., 2001).

A key point here is that, although the relative amount of color processing certainly varies across occipital regions, with area V4 having color processing as its major function, the processing of color-related information does more than simply allow us to tell red from green. The appreciation of color also enriches our capacity to detect motion, depth, and position. In the absence of significant color analysis, dogs and cats thus not only see an essentially black-and-white world, but have reduced visual capacities more generally as well.

An example of the advantage of color vision can be seen in the type of photoreceptors in primates. Sumner and Mollon found that the color system of primates is optimized for differentiating edible fruits from a background of leaves. This ability to differentiate is an important advantage when having to select edible fruits from a complex scene and is especially important when the fruits are partly occluded by leaves, which is fairly common. In fact, color provides important information for object recognition in such cases. A partly occluded yellow banana is quickly seen, whereas a gray banana would be difficult to detect in a black-and-white scene.

Connections of the Visual Cortex

By the late 1960s, the consensus was that the visual cortex is hierarchically organized, with visual information proceeding from area 17 to area 18 to area 19. Each visual area was thought to provide some sort of elaboration on the processing of the preceding area. This strictly hierarchical view is now considered too simple, and has been replaced by the notion of a distributed hierarchical process with multiple parallel and interconnecting pathways at each level, much as illustrated in Figure 10.16.

There is still a hierarchy for vision, but with separate functions. As indicated in Chapter 10, the details of all the connections between the occipital areas and from them to the parietal, temporal, and frontal regions are bewildering, but it is possible to extract a few simple principles (Figure 13.5):

1. V1 (the striate cortex) is the primary vision area: it receives the largest input from the lateral geniculate nucleus of the thalamus and it projects to all other occipital regions. V1 is the first processing level in the hierarchy.

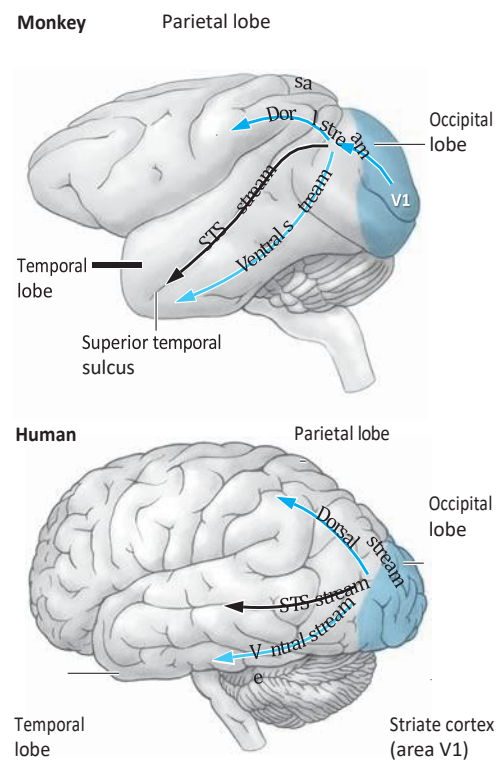


Figure 13.5 Streams of visual processing. The occipitoparietal (dorsal) stream takes part in visual action and flows from area V1 to the posterior parietal visual areas. The occipitotemporal (ventral) stream takes part in object recognition and flows from area V1 to the temporal visual areas. The superior temporal sulcus (STS) stream, where information to and from the dorsal and ventral streams converges, flows from area V1 into the superior temporal sulcus.

2. V2 also projects to all other occipital regions. V2 is the second level.
3. After V2, three distinct parallel pathways emerge en route to the parietal cortex, superior temporal sulcus, and inferior temporal cortex, for further processing.

As we shall see in more detail shortly, the parietal pathway, or **dorsal stream,** has a role in the visual guidance of movement, and the inferior temporal pathway, or **ventral stream,** is concerned with object perception (including color). The middle pathway along the superior temporal sulcus is probably important in visuospatial functions.

A Theory of Occipital-Lobe Function

We have seen that areas V1 and V2 are functionally heterogeneous and that both segregate processing for color, form, and motion. The heterogeneous functions of areas V1 and V2 contrast with the functions of the areas that follow in the hierarchy. In a sense, areas V1 and V2 appear to serve as little mailboxes into which different types of information are assembled before being sent on to the more specialized visual areas.

From areas V1 and V2 flow three parallel pathways that convey different attributes of vision. The information derived from the blob areas of area V1 goes to area V4, considered to be a color area. Cells in area V4 are not solely responsive to color, however; some cells respond to both form and color.

Other information from area V1 (the magnocellular input discussed in Chapter 8) also goes to area V2 and then to area V5, which is specialized to detect motion. Finally, an input from areas V1 and V2 to area V3 is concerned with what Zeki calls "dynamic form"—that is, the shape of objects in motion. Thus, we see that vision begins in the primary cortex (V1), which has multiple functions, and then continues in more specialized zones.

It is not surprising to discover that selective lesions up the hierarchy in areas V3, V4, and V5 produce specific deficits in visual processing. People who suffer damage to area V4 are able to see only in shades of gray. Curiously, patients not only fail to perceive colors but also fail to recall colors from before their injuries or even to imagine colors. In a real sense, the loss of area V4 results in the loss of color cognition, or the ability to think about color. Similarly, a lesion in area V5 produces an inability to perceive objects in motion. Objects at rest are perceived but, when the objects begin to move, they vanish. In principle, a lesion in area V3 will affect form perception but, because area V4 also processes form, a rather large lesion of both V3 and V4 would be required to eliminate form perception.

An important constraint on the functions of areas V3, V4, and V5 is that all these areas receive major input from area V1. People like Colonel P. M., with lesions in area V1, act as though they are blind, but visual input can still get through to higher levels—partly through small projections of the lateral geniculate nucleus to area V2 and partly through projections from the colliculus to the thalamus (the pulvinar) to the cortex. (This system is the tectopulvinar pathway described in Chapter 8.)

People with V1 lesions seem not to be aware of visual input and can be shown to have some aspects of vision only by special testing. Thus, when asked what they see, patients with V1 damage often reply that they see nothing. Nonetheless, they can act on visual information, indicating that they do indeed "see."

Area V1 thus appears to be primary for vision in yet another sense: V1 must function for the brain to make sense out of what the more specialized visual areas are processing. We must note, however, reports of people with significant V1 damage who are capable of some awareness of visual information, such as motion. Barbur and colleagues suggested that the integrity of area V3 may allow this conscious awareness, but this suggestion remains a hypothesis.

Visual Functions Beyond the Occipital Lobe

Neuroscientists have known since the early 1900s that the occipital lobes house vision, but only in the past two decades have they begun to understand the extent of visual processing that takes place beyond the occipital lobes. In fact, it is now clear that more cortex is concerned with vision than with any other function in the primate brain.

Felleman and van Essen's flattened cortical map in Figure 10.17 illustrates that, of the 32 cortical areas (of a total of about 70 in their scheme) that have visual functions in the monkey brain, only 9 are actually in the occipital lobe. The total surface area of the vision-related regions is about 55% of the whole cortical surface, which compares with 11% and 3% for the somatosensory and auditory regions, respectively. (It is interesting that so little of the monkey cortex represents audition, which is certainly evidence of a major difference between the brains of humans and those of monkeys; we humans have a much larger auditory representation, which is no doubt responsible for our preoccupation with both language and music.)

Visual processing in humans therefore does not culminate in secondary visual areas such as areas V3, V4, and V5 but continues within multiple visual regions in the parietal, temporal, and frontal lobes (see Figure 10.17). Functions have not been assigned to all these additional visual regions, but it is possible to speculate on what their functions must be. To do so, we can divide visual phenomena into five general categories: vision for action, action for vision, visual recognition, visual space, and visual attention.

Vision for Action

This category is visual processing required to direct specific movements. For example, when reaching for a particular object such as a cup, the fingers form a specific pattern that allows grasping of the cup. This movement is obviously guided by vision, because people do not need to shape their hands consciously as they reach.

In addition to that for grasping, there must be visual areas that guide all kinds of specific movements, including those of the eyes, head, and whole body. A single system could not easily guide all movements, because the requirements are so different. Reaching to pick up a jellybean requires a very different kind of motor control than that required to duck from a snowball, but both are visually guided.

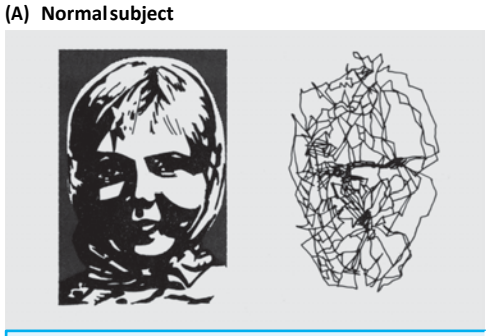


Figure 13.6 Eye movements during the examination of a visual stimulus. (A) The concentration of eye movements (by a normal subject) to distinctive features of the face (eyes, nose, mouth); these movements are directed more to the left side of the photograph. (B) The eye movements of a normal subject examining a sphere (left) and a bust (right). (C) The eye movements of an agnosic subject examining the same shapes. Note the random movements of the agnosic subject. (From A. R. Luria, *The Working Brain*, © 1973, The Copyright Agency of the USSR. Reprinted with permission.)

Finally, vision for action must be sensitive to movement of the target. Catching a moving ball requires specific information about the location, trajectory, speed, and shape of the object. Vision for action is thought to be a function of the parietal visual areas.

Action for Vision

In a more "top down" process, the viewer actively searches for only part of the target object and attends selectively it (Figure 13.6). When we look at a visual stimulus, we do not simply stare at it; rather, we scan the stimulus with numerous eye movements. These movements are not random but tend to focus on important or distinct features of the stimulus.

When we scan a face, we make a lot of eye movements directed toward the eyes and mouth. Curiously, we also make more eye scans directed to the left visual field (the right side of the person's face) than to the right visual field. This scanning bias may be important in the way that we process faces, because it is not found in the scanning of other stimuli. People with deficits in action for vision are likely to have significant deficits in visual perception, although such deficits have not been studied systematically.

An interesting aspect of action for vision consists of the eye movements that we often make when we visualize information. For example, when people are asked to rotate objects mentally in order to answer simple questions about the objects' appearance, they usually make many eye movements, especially to the left. When people are doing things in the dark, such as winding photographic film onto spools for processing, they also make many eye movements. Curiously, if the eyes are closed, these movements stop. Indeed, it appears that it is easier to do many tasks in the dark if the eyes are closed. Because things are done by touch in the dark, the visual system may interfere until the eyes are closed.

Visual Recognition

We enjoy the ability both to recognize objects and to respond to visual information. For example, we can both recognize specific faces and discriminate and interpret different expressions in those faces. Similarly, we can recognize letters or symbols and assign meaning to them.

We can recognize different foods, tools, or body parts, but it is not reasonable to expect that we have different visual regions for each category or object. We may have at least some specialized areas for biologically significant information, such as faces, however. Cells in the temporal cortex appear to be highly specific in their preference for particular faces or hands. These visual areas in the temporal lobe are specialized for visual recognition.

Visual Space

Visual information comes from specific locations in space. This information allows us to direct our movements to objects in space and to assign meaning to objects. But spatial location is not a unitary characteristic. Objects have location both relative to an individual (**egocentric space**) and relative to one another (**allocentric space**).

Egocentric visual space is central to the control of your actions toward objects. It therefore seems likely that visual space is coded in neural systems related to vision for action. In contrast, allocentric properties of objects are necessary for you to construct a memory of spatial location.

A key feature of allocentric spatial coding is that it depends on the identity of particular features of the world. Thus, it is likely to be associated with the regions of visual recognition. In summary, different aspects of spatial processing probably take place in both the parietal and the temporal visual regions, and respective functions are integrated in areas that interact and exchange information.

Visual Attention

We cannot possibly process all the visual information available. This page has shape, color, texture, location, and so on, but the only really important characteristic is that it has words. Thus, when we read the page, we select a specific aspect of visual input and attend to it selectively.

In fact, neurons in the cortex show various attentional mechanisms. For example, neurons may respond selectively to stimuli in particular places or at particular times or if a particular movement is to be executed. Independent mechanisms of attention are probably required both for the guidance of movements (in the parietal lobe) and for object recognition (in the temporal lobe).

Visual Pathways Beyond the Occipital Lobe

Vision evolved first for motion, not for recognition. Simple organisms can detect light and move to or from the light. For example, the single-cell organism *Euglena* alters its swimming pattern as a function of the ambient light levels in different parts of the pond in which it lives. Because sunlight helps manufacture food in this aquatic environment, it is an advantage for *Euglena* to move toward the light.

Notice that *Euglena* need not "perceive" the light or make an internal map of the outside world. Rather, it is only necessary that some type of link exist between the amount of ambient light and locomotion. For *Euglena,* "vision" acts to guide movement—the most primitive form of vision for action.

However, our vision is far more complicated than that of *Euglena.* But even much of human vision can be understood without reference to recognition. Consider, for example, a major-league baseball batter who swings at a ball before it is possible for him to perceive what the object actually is. The visual guidance of his movement is independent of his recognition of the ball.

Nonetheless, as primitive animals interacted with their environment, they are adapted to learn more about their environment. Thus distinct visual systems evolved to recognize objects in the environment. The system of knowing what an object is includes the flow of visual information from area V1 to

the temporal lobe, called the ventral stream (see Figure 13.5). The system controlling the visual guidance of movements includes the flow of information from area V1 to the parietal lobe, known as the dorsal stream.

The distinction between the ventral and the dorsal streams can be seen clearly in a series of patients studied by Milner and Goodale. They first described D. F., a patient who was blind but who nevertheless shaped her hand appropriately when asked to reach for objects. Her dorsal stream was intact, as revealed by the fact that she could "unconsciously" see location, size, and shape. On the other hand, Milner and Goodale noted that patients with dorsal-stream damage consciously reported seeing objects but could not reach accurately or shape the hand appropriately when reaching.

Milner and Goodale proposed that the dorsal stream should be thought of as a set of systems for the on-line visual control of action. Their argument is based on three main lines of evidence:

1. The predominant characteristic of the neurons in posterior parietal regions is that they are active during a combination of visual stimulation and associated behavior. For example, cells may be active only when a monkey reaches out to a particular object. Looking at an object in the absence of movement does not activate the neurons. Thus, these "visual" neurons are unique in that they are active only when the brain acts on visual information.

2. These posterior parietal neurons can therefore be characterized as an interface between analysis of the visual world and motor action taken on it. The demands of action have important implications for what type of information must be sent to the parietal cortex—information such as object shape, movement, and location. Each of these visual features is likely to be coded separately, and at least three distinct pathways within the dorsal stream run from area V1 to the parietal cortex. As illustrated on the right in Figure 13.7, one pathway goes from area V1 directly to area V5 to parietal cortex, a second goes from area V1 to areas V5 and V3a and then to parietal regions, and a third goes from area V1 to area V2 to the parietal cortex. These three pathways must certainly be functionally dissociable.

3. Most of the visual impairments associated with lesions to the parietal cortex can be characterized as visuomotor or orientational. (We return to this subject in Chapter 14.)

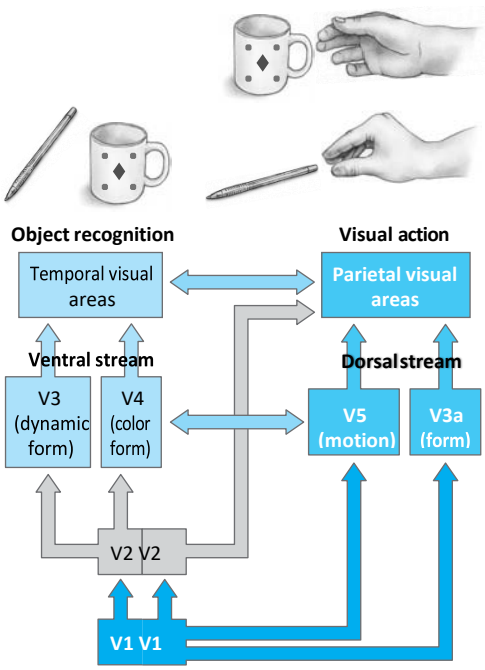


Figure 13.7 A summary of the visual processing hierarchy. The dorsal stream, which takes part in visual action, guides movements such as the hand postures for grasping a mug or pen, as illustrated. The ventral stream, which takes part in object recognition, identifies objects such as mugs and pens in our visual world. The dorsal and ventral streams exchange information through polysensory neurons in the superior temporal sulcus stream, as shown by double-headed arrows. (After Goodale, 1993.)

The Milner-Goodale model is an important theoretical advance in understanding how our visual brain is organized. As detailed in Figure 13.7, two distinct visual streams have evolved to use visual information in two fundamentally different ways: the dorsal stream for guiding movements and the ventral for identifying objects. This model can likely be applied to the organization of the auditory and somatosensory systems as well—both systems also function to guide movements and identify stimuli. An important point here is that we are conscious of only a small amount of what the brain actually does; even with effort, we cannot gain awareness of much of our sensory processing.

One wrinkle must be added to the Milner-Goodale model—the third stream of visual processing, which originates from

structures associated with both the parietal and the temporal pathways and flows to a region of the temporal lobe that is buried in the superior temporal sulcus (see Figure 13.5). The superior temporal sulcus is characterized by **polysensory neurons**—neurons that are responsive to both visual and auditory or both visual and somatosensory input. The interaction of the parietal and temporal streams in the superior temporal sulcus is probably due to interaction between the dorsal and the ventral—the "action" and "recognition"—streams.

Imaging Studies of Dorsal and Ventral Streams

Brain regions associated with specific visual pathways can be identified by measuring regional blood flow as people perform visual tasks. Ungerleider and Haxby reviewed such PET studies, as summarized in Figure 13.8.

In studies by Haxby and colleagues, subjects were given two tasks. In the first, the subjects indicated which of two faces was identical with a sample face. In the second, the subjects were asked to identify which of two stimuli had a dot (or square) in the same location as in a sample. The results showed activation of the temporal regions for the facial stimuli and activation of the posterior parietal region for the location task (see Figure 13.8A). Note, in addition, the activation of frontal areas for the spatial task, supporting the idea that the frontal lobe plays a role in certain aspects of visual processing.

One difficulty with interpretation of the spatial-task PET images is that subjects have to make eye movements, which activate regions in the dorsal stream; so whether the spatial or the movement components activate the parietal region is not clear. The important point, however, is that different regions take part in the two tasks.

A similar dissociation was identified among the processes that detect motion, color, and shape (see Figure 13.8B). Detection of motion activates regions in the

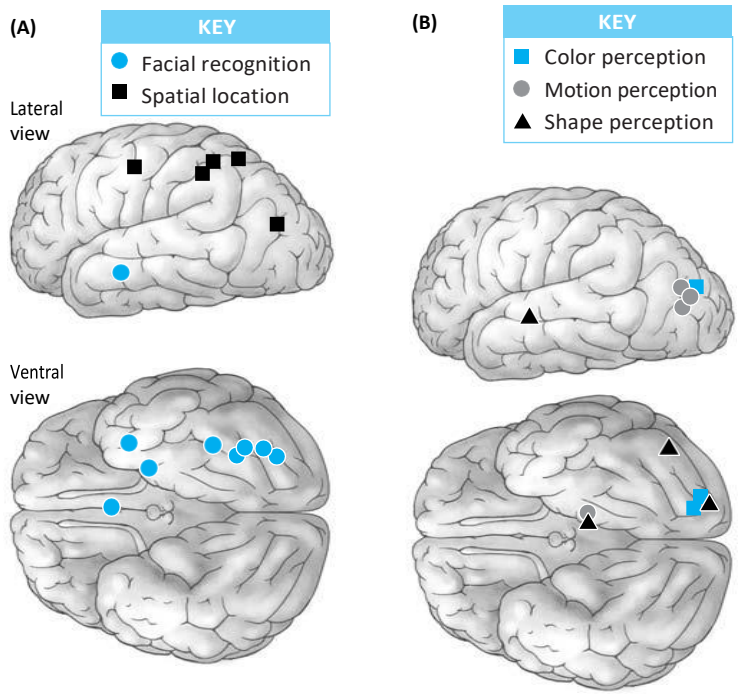


Figure 13.8 Summary of results of PET studies illustrating selective activation of (A) cortical regions by tasks of facial recognition (circles) versus spatial location (squares) and (B) areas associated with perception of color (squares), motion (circles), and shape (triangles). (After Ungerleider and Haxby, 1994.)

vicinity of area V5, whereas detection of shape activates regions along the superior temporal sulcus and the ventral region of the temporal lobe. The perception of color is associated with activation of the region of the lingual gyrus, which is the location of area V4. One study also found activation of a lateral occipital region, which is difficult to interpret in the light of lesion studies. This study made special visual attention demands of its participants, potentially an important factor in interpreting the observed activation.

In summary, studies of regional blood flow in normal subjects show results consistent with the general notion of two separate visual streams, one to the parietal lobe and the other to the temporal lobe. In addition, it is clear that separate visual functions reside in different temporo-occipital regions.

Disorders of Visual Pathways

Before we consider the deficits associated with damage to the visual pathways, we must revisit two key elements in the way in which the brain organizes the visual fields:

1. The left half of each retina sends its projections to the right side of the brain, whereas the right half of each retina sends its projections to the left side of the brain (see Figure 11.5). Thus, the representation of each side of the visual world seen by each eye is sent to the same place in area V1. As a result, damage to area V1 affects vision in both eyes. Conversely, if a visual disturbance is restricted to just one eye, then the damage must be outside the brain, either in the retina or in the optic nerve.
2. Different parts of the visual field are topographically represented in different parts of area V1 (Figure 13.9). Thus, injury to a specific region of area V1 produces a loss of vision in a specific part of the visual world.

Now let us consider what happens when there is damage to different places in the visual pathways, as keyed on Figure 13.9.

Destruction of the retina or optic nerve of one eye produces monocular blindness—the loss of sight in that eye. A lesion of the medial region of the optic chiasm severs the crossing fibers, producing **bitemporal hemianopia**—loss of vision of both temporal fields. This symptom can arise when a tumor develops in the pituitary gland, which sits medially, next to the chiasm. As the tumor grows, it can put pressure on the medial part of the chiasm and produce the loss, or disturbance, of lateral vision.

A lesion of the lateral chiasm results in a loss of vision of one nasal field, or *nasal hemianopia*. Complete cuts of the optic tract, lateral geniculate body, or area V1 result in **homonymous hemianopia**—blindness of one entire visual field (see Figures 13.9 and 13.10A). Note that, because the disturbance affects information coming from both eyes, the visual defect is present in both eyes.

Indeed, the effects of such injuries enable investigators to determine whether a lesion is in the eye or optic tract versus the optic nerve or brain. The former injuries produce visual disturbance in one eye, whereas the latter injuries produce visual disturbance in the visual field and thus in both eyes. Should this lesion be partial, as is often the case, only a part (quadrant) of the visual field is destroyed (see Figures 13.9 and 13.10).

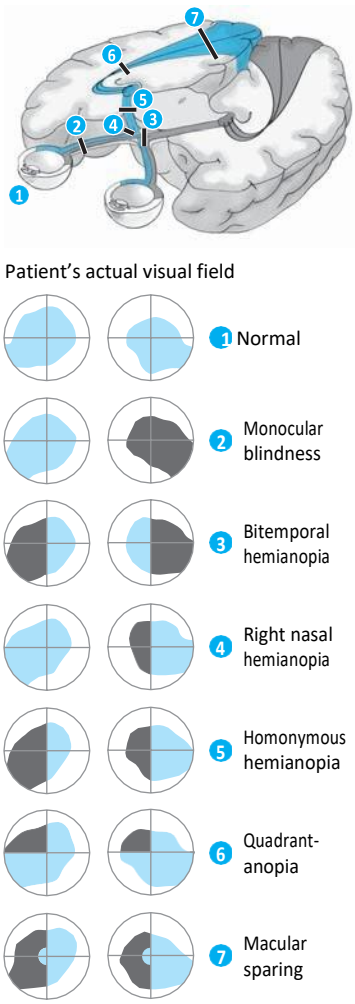


Figure 13.9 Visual defects subsequent to damage at different levels of the visual system, keyed by number. A darkened region in the visual field denotes a blind area. (After Curtis, 1972.)

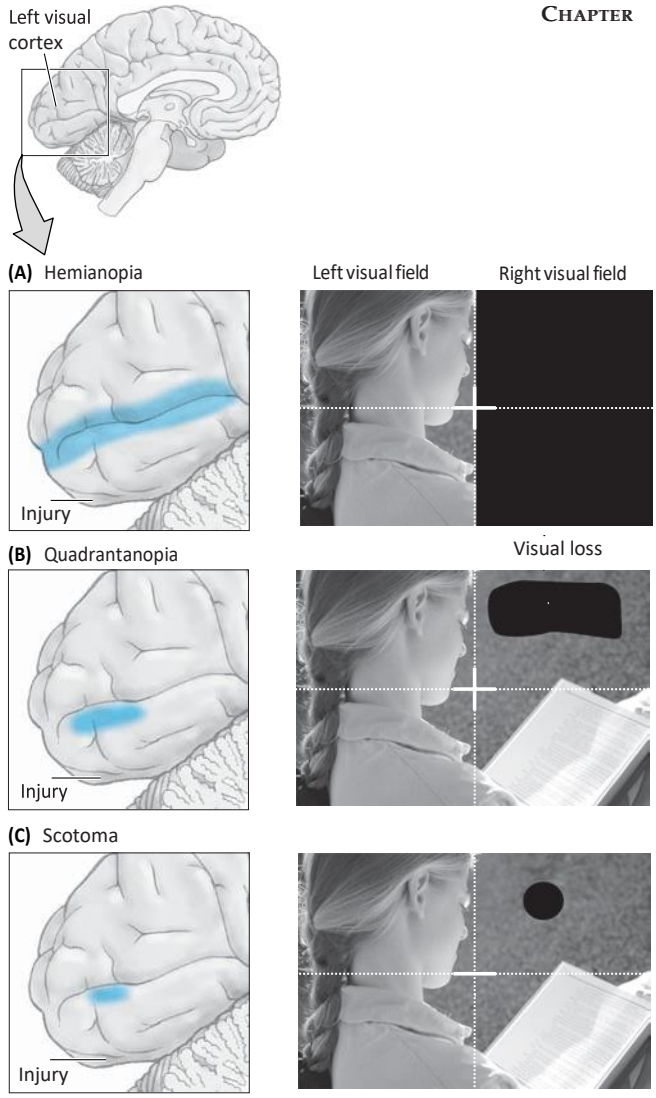


Figure 13.10 Consequences of lesions in area V1. The blue-dark areas indicate the regions of visual loss. (A) The effect of a complete lesion of area V1 in the left hemisphere is hemianopia affecting the right visual field. (B) A large lesion of the lower lip of the calcarine fissure produces a quadrantanopia that affects most of the upper-right visual quadrant. (C) A smaller lesion of the lower lip of the calcarine fissure results in a smaller injury, a scotoma. (Jim Pickerell/Stock Connection/PictureQuest.)

Lesions of the occipital lobe often spare the central, or macular, region of the visual field, although the reason is uncertain. The most reasonable explanations are that (1) the macular region receives a double vascular supply, from both the middle and the posterior cerebral arteries, making it more resilient to large hemispheric lesions, or (2) the foveal region of the retina projects to both hemispheres, and so, even if one occipital lobe is destroyed, the other receives projections from the fovea. The first explanation is more likely.

Macular sparing of the central visual field helps to differentiate lesions of the optic tract or thalamus from cortical lesions, because macular sparing occurs only after lesions (usually large) to the visual cortex. Macular sparing does not always occur, however, and many people with visual-cortex lesions have a complete loss of vision in one-quarter (quadrantanopia) or one-half (hemianopia) of the fovea (see Figures 13.9 and 13.10B). A curious aspect of both hemianopia and quadrantanopia is that the border between the impaired visual area and the adjacent, intact visual field, or quadrant, is sharp, much as if a pair of scissors were used to cut away part of the visual field (see Figure 13.9). This sharp demarcation of intact and impaired visual regions is due to the anatomical segregation between the left and the right and the upper and the lower visual fields.

Small lesions of the occipital lobe often produce **scotomas,** small blind spots in the visual field (Figure 13.10C). A curious aspect of scotomas is that people are often totally unaware of them because of *nystagmus* (constant, tiny, involuntary eye movements) and "spontaneous filling in" by the visual system. The eyes are usually in constant motion; so the scotoma moves about the visual field, allowing the brain to perceive all the information in the field. If the eyes are held still, the visual system actually completes objects, faces, and so on, resulting in a normal percept of the stimulus.

The visual system may cover up the scotoma so successfully that its presence can be demonstrated to the patient only by "tricking" the visual system. Such tricking can be achieved by placing objects entirely within the scotoma region of the patient's visual field and, without allowing the patient to shift gaze, asking what the object is. If no object is reported, the examiner moves the object out of the scotoma so that it suddenly "appears" in the intact region of the patient's visual field, thus demonstrating the existence of a blind region.

A similar phenomenon can be demonstrated in your own "blind spot." Stand beside a table, close or cover one eye, stare at a spot on the table, and move a pencil along the table laterally, from directly below your nose to between 20 and 30 cm toward the periphery. Part of the pencil will vanish when you reach the blind spot. You can move the pencil through the blind spot slowly, and it will suddenly reappear on the other side. Notice that, like a scotoma, the normal blind spot is not noticeable, even when you look around the world with just one eye. Even the normal brain "fills in" missing bits of the visual world.

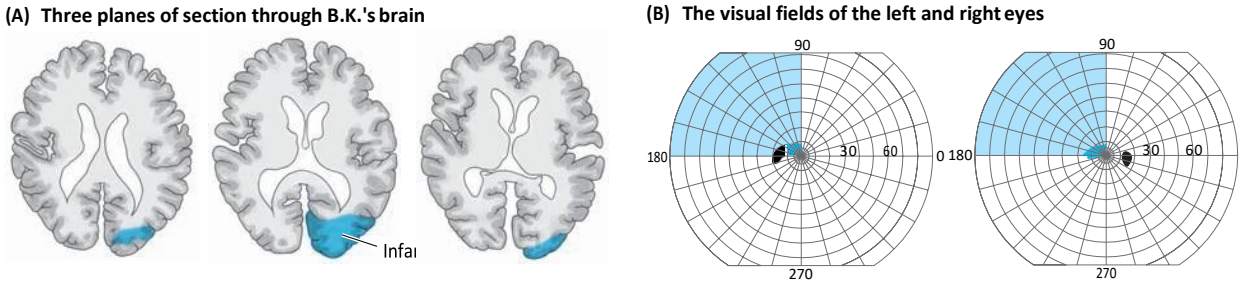
Disorders of Cortical Function

Research into selective disturbances of human visual functions is limited mainly to case studies, such as case P. M., whom you met at the beginning of this chapter, and these natural lesions seldom respect the boundaries of specific visual areas. Several case histories, each with distinctly different symptoms and pathology, will give you a feeling for the specific symptoms of injury to the visual cortex. We begin with damage to area V1 and proceed along the hierarchy to higher areas and more-complicated visual disturbances.

Case B. K.: V1 Damage and a Scotoma

One morning B. K. awoke to discover that he was hemianopic in the left visual field. Given a history of classic migraine, in which the aura was nearly always in the left visual field, it is likely that he had a migraine stroke. Within a few hours, the left lower field began to return, but the left upper quadrant was slow to show any change.

A CT scan (Figure 13.11A) showed a clear infarct (dead tissue) in the right occipital area. The size of a visual-field defect is routinely measured with perimetry, a standardized method in which the subject fixates on a black dot in the center of a large white hemisphere. A small light is moved around the field, and the task is to indicate when it can be seen. The brightness and size of the light can be varied, thus manipulating the difficulty of the task.



Performance is mapped by indicating the area of "blindness" on a schematic map of the visual fields (Figure 13.11B). Size in the visual field is measured by visual angle, in degrees. (A degree is roughly the size of your thumb viewed at arm's length.) Thus, for B. K., the area of complete inability to perceive even a very large bright light is measured from the center, 6° upward along the vertical midline and about 15° laterally along the horizontal midline. The area beyond this zone in the left upper quadrant does not have normal vision, however, because B. K. is still unable to perceive less-bright lights in this area.

Figure 13.11 Scan and map of B. K.'s brain. (A) Schematic representation of B. K.'s CT scan, showing the infarct in the right occipital area in different views. (B) Map of B. K.'s visual fields 6 months after the stroke. Subnormal vision persists in the upper-left quadrant.

The nature of B. K.'s visual defects can be illustrated best in the context of their poststroke evolution. For the first 2 to 3 days, his visual field appeared dark, much as though a piece of smoked glass were blocking his view of the world beyond. On the fourth day, this darkness had disappeared and was replaced by "visual noise" (a scintillating scotoma) throughout much of the field, especially in the area of the scotoma. Visual noise is best described as being like colored "snow" on a television screen. At about the same time, B. K. first perceived movement in the field as a traveling "wave," much like ripples on a pond. There was no perception of form or pattern.

A curious phenomenon was first observed during perimetry testing 4 days after the stroke. If the stimulus light was moved into the blind field, it was not perceived until it moved into another quadrant. Curiously, however, B. K. immediately became aware (in hindsight) that the light had been present in the blind field and could accurately state where it entered the blind field. In other words, B. K. perceived location without being able to perceive content. Recall that Colonel P. M also experienced this phenomenon, known as **blindsight.**

In the ensuing 4 to 6 months, the area of blindness decreased somewhat and acuity in the periphery improved significantly for B. K. Nonetheless, roughly 15 years later, form vision remains poor in the left upper quadrant, outside the scotoma. The scintillating colored snow is still present, showing little change from the first few days after the stroke.

The visual phenomena observed by B. K. indicate that area V1 (and perhaps area V2) probably has an area of total cell death (the dense scotoma). The presence of poor form vision in the rest of the quadrant may be due to a loss of some but not all neurons in area V1, possibly only those neurons that are especially sensitive to a period of reduced blood flow, known as ischemia. The poor form vision might also be attributed to the fact that other visual areas, especially area V2, remain intact.

B. K.'s symptoms show that other occipital areas are functional, because he perceives color and motion even though there is no form perception. Thus, B. K. can accurately perceive the color or motion of objects that he cannot

identify. Those who are myopic (nearsighted) experience a similar phenomenon: the colors of objects or lights can be perceived, whereas the form is not recognizable. B. K.'s stroke thus indicates the presence of at least four independent visual functions: form (which is absent), as well as color, movement, and location (which are spared).

The loss of one-quarter of the fovea leads B. K. to make a variety of visual errors. Immediately after the stroke, he was able to read only with great difficulty. When we look at a word, the fixation point is in the center of the word; so, for B. K., half of the word is absent. Indeed, he had difficulty finding the edge of the page because it was in the blind field. Normal reading returned as B. K. learned to direct his gaze slightly to the left and upward (probably about 2° in each direction), which allowed words to fall in the normal visual field.

This "recovery" took about 6 weeks. Returning to playing squash and tennis was equally challenging because, when a ball entered the scotoma, it was lost. Similarly, facial recognition was slower than it had been before the stroke, because the information in the left visual field appears to be particularly important for face recognition.

Case D. B.: V1 Damage and Blindsight

D. B. is one of the most extensively studied people with visual disturbance from an occipital lesion (see the detailed monograph by Weiskrantz). D. B.'s right calcarine fissure was removed surgically to excise an angioma, which is a collection of abnormal blood vessels. D. B. therefore has a hemianopia based on standard perimetry but nevertheless has surprising visual capacities.

When questioned about his vision in the left field, D. B. usually reports that he sees nothing, as did P. M. and B. K. Occasionally, D. B. indicates that he had a "feeling" that a stimulus was "approaching" or was "smooth" or "jagged." But, according to Weiskrantz, D. B. always stresses that he "saw" nothing, that typically he is guessing, and that he is at a loss for words to describe any conscious perception.

In contrast, when D. B. was asked to point to locations in the impaired field in which spots of light were turned on briefly, he was surprisingly accurate. His blindsight contrasts with his subjective impression that he saw nothing at all. Furthermore, he appears to be able to discriminate the orientation of lines, which he could not report "seeing." Thus, he can discriminate a 10° difference in orientation between two successively presented gratings in his impaired field.

Finally, D. B. can detect some forms of movement. When a vigorously moving stimulus was used, he reported "seeing" something. In this case, he did not report actually seeing a visual stimulus but rather spoke of complex patterns of lines and grids. These patterns may have been something like B. K.'s moving lines. In summary, D. B. has "cortical blindness," or blindsight, in which he reports no conscious awareness of "seeing" but still is able to report the movement and location of objects that he cannot recognize.

Case J. I.: V4 Damage and Color

Oliver Sacks and Robert Wasserman report the touching story of J. I., an artist who suddenly became color-blind. In 1986, the man was in a car accident in which he sustained a concussion. His principal symptoms after

the injury were an inability to distinguish any colors whatsoever, but his visual acuity had actually improved. "Within days . . . my vision was that of an eagle—I can see a worm wiggling a block away. The sharpness of focus is incredible."

The effect of the loss of color vision was far greater than one would have expected. J. I. could barely stand the pain of living in a world that appeared in shades of gray. He found the changed appearance of people unbearable, because their flesh was an abhorrent gray ("rat-colored") to him. He found foods disgusting in their grayish, dead appearance, and he had to close his eyes to eat. He could not even imagine colors any longer. The mental image of a tomato looked as black as its actual appearance. Even his dreams, which had once been in vivid colors, were now in black and gray.

Detailed visual testing by Sacks and Wasserman, and later by Zeki, revealed that J. I. was color-blind by the usual definitions, but this color blindness was attributed to specific damage to the occipital cortex. In addition, it did appear that his acuity had improved, especially at twilight or at night. Two years after his injury, J. I.'s despair had declined, and he appeared to no longer be able to remember color well. This failure to remember color is curious, because people who become blind through injury to the eyes or optic nerves do not lose their imagery or memory of color. There is little doubt from J. I.'s case that imagery and memory rely on the operation of at least some cortical structures necessary for the original perception.

Case P. B.: Conscious Color Perception in a Blind Patient

Zeki and his colleagues described the case of a man who was electrocuted, resulting in cardiac and respiratory arrest. P. B. was resuscitated but had suffered brain ischemia that produced a large area of posterior cortical damage. P. B. was left virtually blind, although he can detect the presence or absence of light. The remarkable visual feature, however, is that P. B.'s capacity to identify and name colors remains intact, as does his ability to name the typical color of imagined objects.

P. B.'s vision is in many ways opposite that of J. I.; the results of fMRI studies show that P. B. has activation in areas V1 and V2 in response to colored stimuli. As we reflect on the visual capacity of P. B., it is hard to imagine a world that is filled with color but no form, almost like an out-of-focus kaleidoscope that changes as we gaze around the world.

Case L. M.: V5 Damage and the Perception of Movement

Zihl and his colleagues reported the case of a 43-year-old woman who had a bilateral posterior injury resulting from a vascular abnormality. Her primary chronic complaint was a loss of movement vision. For example, she had difficulty pouring tea into a cup because the fluid appeared to be frozen. And she could not stop pouring, because she could not see the fluid level rise.

L. M. found being in a room with other people disturbing because she could not see them moving; they suddenly appeared "here or there," but she did not see them move in between. The results of other tests of visual function appeared essentially normal. She could discriminate colors, recognize objects, and read and write.

Her condition is especially intriguing because we would not believe intuitively that such a syndrome is likely. Loss of color or form vision fits with our everyday experience that people can be color-blind or myopic; loss of the ability to see moving objects is counterintuitive indeed. Case L. M. is important because she shows that the brain must analyze movement of form separately from the form itself.

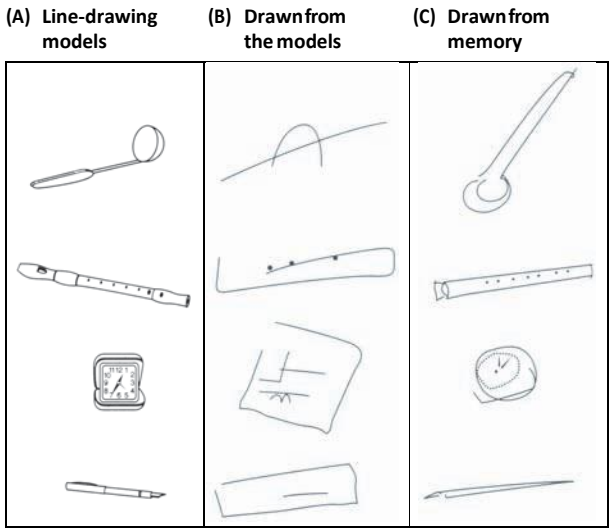


Figure 13.12 Samples of D. F.'s drawings. (A) Examples of the original line drawings presented to D. F. (B) Examples of D. F.'s drawings of the models. (C) D. F.'s drawings based on memory of the models. Note that the drawings from memory are superior to the copies of the line drawings of the models.

Case D. F.: Occipital Damage and Visual Agnosia

Visual agnosia is the term coined by Sigmund Freud for an inability to combine individual visual impressions into complete patterns—thus, the inability to recognize objects or their pictorial representations or the inability to draw or copy them. Goodale and Milner and their colleagues have extensively studied a 35-year-old visual agnosic who suffered carbon monoxide poisoning that resulted in bilateral damage to the lateral occipital region (mainly areas 18 and 19) and in the tissue at the junction of the parietal and occipital cortex.

C. F., whom we met earlier in the chapter in considering Goodale and Milner's distinction between the dorsal and the ventral streams, has essentially normal color vision and can see well enough to get around in the world. Her principal deficit is *visual-form agnosia*, a severe inability to recognize line drawings of objects. Thus, although D. F. can recognize many actual objects, she is unable to recognize drawings of them. Furthermore, as illustrated in Figure 13.12, although she can draw objects from memory, she has real difficulty in drawing objects from life and even more difficulty in copying line drawings. Thus, D. F. appears to have a serious defect in form perception.

Recall that the remarkable thing about D. F. is her apparently nearly intact ability to guide hand and finger movements toward objects that she cannot recognize. For example, although D. F. had a gross deficit in judging lines as horizontal or vertical, she could reach out and "post" a hand-held card into a slot rotated to different orientations, as illustrated in Figure 13.13. Indeed, analysis of video records of D. F.'s reaching revealed that, like normal control subjects, she began to orient the card correctly even as her hand was being raised

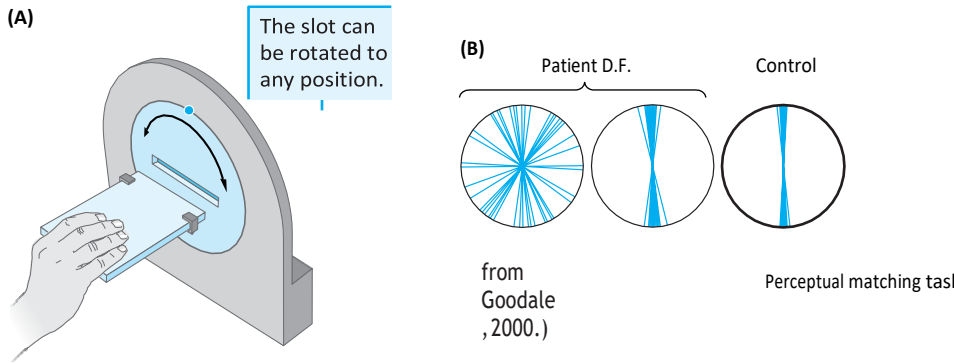


Figure 13.13 Testing visuomotor guidance. (A) The apparatus that was used to test sensitivity to orientation in patient D. F. The slot could be placed in any orientation around the clock. The task is to "post" the card into the slot as shown. (B) Plots of the orientation of the card in a perceptual matching task and in the visuomotor posting task. For illustration, the correct orientation has been rotated to vertical. D. F. was unable to match the orientation of the card to that of the slot unless she made a movement to post it. (Adapted with permission from Goodale, 2000.)

Visuo motor posti ng task

from the start position of the task. In other words, D. F. could use visual form information to guide movements to objects (the dorsal stream), but she could not use visual information to recognize the same objects (the ventral stream).

Case V. K.: Parietal Damage and Visuomotor Guidance

Damage to the posterior parietal lobe produces a disorder known as **optic ataxia,** a deficit in visually guided hand movements, such as reaching, that cannot be ascribed to motor, somatosensory, or visual-field or -acuity deficits. V. K. is a woman with bilateral hemorrhages in the occipitoparietal regions, as described by Jakobson and colleagues. Although V. K. initially appeared virtually blind, her symptoms dissipated in a month, and she was left with disordered control of her gaze, impairment in visual attention, and optic ataxia. (Collectively, these symptoms are known as Balint's syndrome; see Chapter 14.)

V.K. had good form and color vision and could recognize and name objects; however, her ability to reach for objects was grossly impaired. Thus, in contrast with D. F., who was able to reach and orient her hand posture toward different objects that she could not perceive, V. K. was unable to coordinate reaching and grasping for objects that she could perceive.

This difficulty was not merely one of being unable to direct movements in space, because V. K. could point to objects. What she could not do was to form the appropriate hand postures needed to grasp objects of different shapes, as illustrated in Figure 13.14. Taken together, cases D. F. and V. K. suggest that the mechanisms underlying the conscious perception of object form are dissociable from the mechanisms controlling visually guided movements to the same objects.

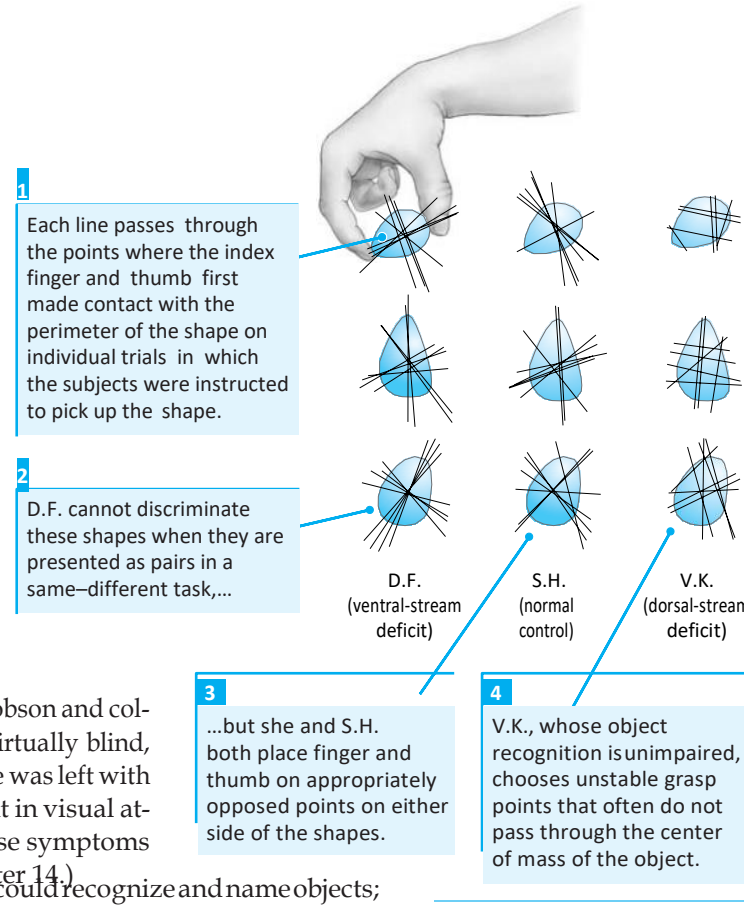


Figure 13.14 The brain has different systems for visual object recognition and visual guidance of movement. Representative "grasping" axes for three different shapes by patient D. F. with visual form agnosia (ventral-stream deficit), by control subject S. H. with no brain damage and by V. K., a patient with bilateral occipitoparietal damage resulting in optic ataxia (dorsal-stream deficit). Even though D. F. does not recognize the object, she perceives enough information about shape to control her grasp as she picks it up. In contrast, V. K. recognizes objects but cannot control her movements in relation to them. (Adapted from Milner and Goodale, 1995.)

Cases D. and T.: Higher-Level Visual Processes

Two cases described by Campbell and colleagues illustrate an intriguing dissociation of visual functions. Case D. has a right occipitotemporal lesion associated with a left upper quadrantanopia that extends into the lower quadrant. As would be expected from B. K.'s case, D. had some initial difficulties in reading, but her language abilities were intact. Curiously, she was completely unable to recognize people by their faces and had difficulty identifying handwriting, including her own.

Recall from the case presented at the beginning of this chapter that P.M. also had difficulty in recognizing faces. His view on his difficulty was that, although he could see the different bits of the face quite clearly, he had trouble putting it all together because, unless a person was a long way off, the entire face was not in the visual field all at once. You can imagine what it would be like to try to recognize people by looking at snapshots of different parts of their faces.

The facial-recognition deficit, **prosopagnosia,** is particularly interesting because many prosopagnosics cannot recognize even their own faces in a mirror. Although D. could not recognize faces, she could make use of information in faces. For example, when given various tests of lip reading, she appeared completely normal. Furthermore, she could imitate the facial movements and expressions of another person.

Case T. provides an interesting contrast to case D. Case T. has a left occipitotemporal lesion with a right hemianopia. She had great difficulty reading (**alexia**) and was unable to name colors, even though she could discriminate them. In contrast with D., T. had no difficulty in recognizing familiar faces but was impaired in lip reading.

Taken together, cases D. and T. indicate that face identification and the extraction of speech information from faces do not call on the same cortical systems. In addition, the fact that D. has a lesion on the right and a deficit in face identification and that T. has a lesion on the left and a deficit in lip reading suggests a relative asymmetry in some aspects of occipital-lobe functions. Exactly what visual processes are impaired in the two cases and what the necessary lesions for deficits in facial recognition and lip reading might be remain to be shown.

Conclusions from Case Studies

Several conclusions can be extracted from the behavior and pathology of these cases:

- There are clearly distinct syndromes of visual disturbance.
- Some symptoms can be taken as evidence of a fundamental dissociation between vision for guiding movements (the dorsal stream) and visual recognition (the ventral stream).
- The dissociability of the symptoms in the various patients implies that our introspective view of a unified visual experience is false. The fact that objects can be seen when they are still but not when they are moving is particularly disturbing, because it seems to defy the commonsense view that an object is the same object whether it is moving or still. Clearly the brain does not treat objects in the same way in the two conditions.
- Neuroscientists have at least suggestive evidence of an asymmetry in occipital-lobe functions.

Visual Agnosia

A difficulty in describing the symptomatology and pathology of agnosia is the bewildering variety of patients and symptoms discussed in the neurological literature. Another, as Farah has pointed out, is that there is no agreement on a taxonomy of agnosia, which makes classifying different patterns of symptoms very difficult. We shall separate visual agnosias into object agnosias and other agnosias.

Object Agnosias

The traditional way to classify visual-object agnosia is to distinguish two broad forms: apperceptive agnosia and associative agnosia.

Apperceptive Agnosia

Any failure of object recognition in which relatively basic visual functions (acuity, color, motion) are preserved is apperceptive. This agnosia category has been applied to an extremely heterogeneous set of patients, but the fundamental deficit is an inability to develop a percept of the structure of an object or objects. In the simplest case, patients are simply unable to recognize, copy, or match simple shapes, much like case D. F.

Many patients have another unusual symptom, too—often referred to as **simultagnosia.** In this case, patients can perceive the basic shape of an object, but they are unable to perceive more than one object at a time. Thus, if two objects are presented together, only one is perceived. Such patients often act as though they were blind, possibly because they are simply overwhelmed by the task at hand. Imagine trying to see the world one object at a time.

Apperceptive agnosia does not result from a restricted lesion but usually follows gross bilateral damage to the lateral parts of the occipital lobes, including regions sending outputs to the ventral stream. Such injuries are probably most commonly associated with carbon monoxide poisoning, which appears to produce neuronal death in "watershed" regions—that is, regions lying in the border areas between territories of different arterial systems.

Associative Agnosia

The inability to recognize an object despite an apparent perception of the object is associative agnosia. Thus, the associative agnosic can copy a drawing rather accurately, indicating a coherent percept, but cannot identify it. Associative agnosia is therefore conceived as being at a "higher cognitive" level of processing that is associated with stored information about objects—that is, with memory.

In effect, failure of object recognition is a defect in memory that affects not only past knowledge about the object but also the acquisition of new knowledge. Associative agnosias are more likely with damage to regions in the ventral stream that are farther up the processing hierarchy, such as the anterior temporal lobe.

Other Agnosias

A critical point in understanding the nature of visual agnosia is that the most commonly affected region is the tissue at the occipitotemporal border, which is part of the ventral visual pathway. Visual agnosias do not appear to result from damage to the dorsal stream. Note, however, that agnosias are at least partly dissociable, which means that there must be different streams of visual information processing within the ventral pathway. We now briefly consider three other visual agnosias.

Prosopagnosia

Patients with facial agnosia (recall cases D. and P. M.) cannot recognize any previously known faces, including their own as seen in a mirror or photograph. They can recognize people by face information, however, such as a birthmark, mustache, or characteristic hairdo.

Prosopagnosics may not accept the fact that they cannot recognize their own faces, probably because they know who must be in the mirror and thus see themselves. We saw one young woman who was convinced of the severity of her problem only when she was presented with her identical twin sister. When asked who her twin was, she indicated that she had never seen the woman before. Imagine her amazement to discover that the person was her twin sister.

According to Damasio and colleagues, most facial agnosics can tell human from nonhuman faces and can recognize facial expressions normally. All postmortem studies on facial agnosics have found bilateral damage, and the results of imaging studies in living patients confirm the bilateral nature of the injury in most patients, with the damage centered in the region below the calcarine fissure at the temporal junction. These results imply that the process of facial recognition is probably bilateral, but asymmetrical.

Alexia

An inability to read has often been seen as the complementary symptom to facial-recognition deficits. Alexia is most likely to result from damage to the left fusiform and lingual areas. Either hemisphere can read letters, but only the left hemisphere appears able to combine the letters to form lexical representations (that is, words). Alexia can be conceived to be a form of object agnosia in which there is an inability to construct perceptual wholes from parts or to be a form of associative agnosia, in which case word memory (the lexical store) is either damaged or inaccessible.

Visuospatial Agnosia

Among this variety of disorders of spatial perception and orientation, one disruptive form is *topographical disorientation*—the inability to find one's way around familiar environments such as one's neighborhood. People with this deficit seem unable to recognize landmarks that would indicate the appropriate direction in which to travel. Most people with topographical disorientation have other visual deficits, especially defects in facial recognition. Thus, it is not surprising to find that the critical area for this disorder lies in the right medial occipitotemporal region, including the fusiform and lingual gyri.

Why Are Faces Special?

Most of us probably spend more time looking at faces than at any other single stimulus. Infants prefer to look at faces almost from birth, and adults are excellent at identifying familiar faces despite large variations in expression and viewing angles, even when the faces are disguised with beards, spectacles, or hats. Faces also convey a wealth of social information, and we humans are unique among primates in spending a good deal of time looking directly at the faces of other members of our species.

Figure 13.15 The "Thatcher illusion." Look at the face of former British Prime Minister Margaret Thatcher as presented (upside down) and then invert the page and look again. There is a compelling illusion of normalcy in the inverted face but, in the upright view, the reconfigured face appears hideous. Lady Thatcher was the original subject of the illusion that now bears her name.

The importance of faces as visual stimuli has led to the idea that a special pathway exists in the visual system for the analysis of faces (see Farah, 1998, for a review). Several lines of evidence support this view. In the first place, the results of studies of monkeys show neurons in the temporal lobe that are specifically tuned to different faces, with some cells attuned to facial identity and others to facial expression. (We return to these cells in Chapter 15.) In the second place, inverting a photograph of any object that has a usual right side up makes it harder to recognize, but the effect on faces is disproportionate (see a review by Valentine).

Similarly, we are particularly sensitive to the configuration of upright faces. Consider the illusion shown in Figure 13.15, which illustrates this effect. The importance of an upright orientation to facial perception is also seen in imaging studies. For example, Haxby and his colleagues showed that inverted faces are processed by the same cortical regions as are other visual stimuli, whereas upright faces are processed in a separate face-perception system. This face-perception system is surprisingly extensive and includes regions not only in the occipital lobe but also in several different regions of the temporal lobe.

Figure 13.16 summarizes a model by Haxby and colleagues in which different aspects of facial perception (such as facial expression versus identity) are analyzed in core visual areas in the temporal part of the ventral stream. The model also includes other cortical regions as an "extended system" that includes the analysis of other facial characteristics such as emotion and lip reading. The key point here is that the analysis of faces is unlike that of other visual stimuli.

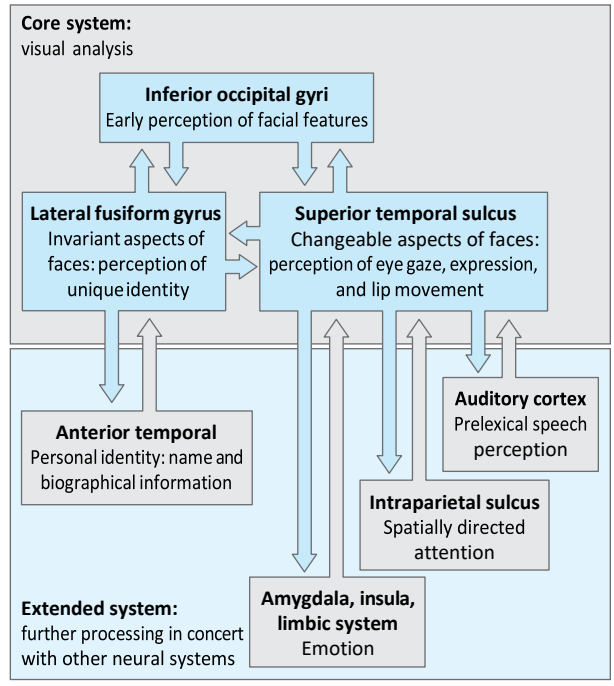


Figure 13.16 A model of the distributed human neural system for face perception. The model is divided into a core system, consisting of occipital and temporal regions, and an extended system, including regions that are part of neural systems for other cognitive functions. The fusiform gyrus is an occipital region lying on the ventral surface of the temporal lobe. (After Haxby, Hoffman, and Gobbini, 2000.)

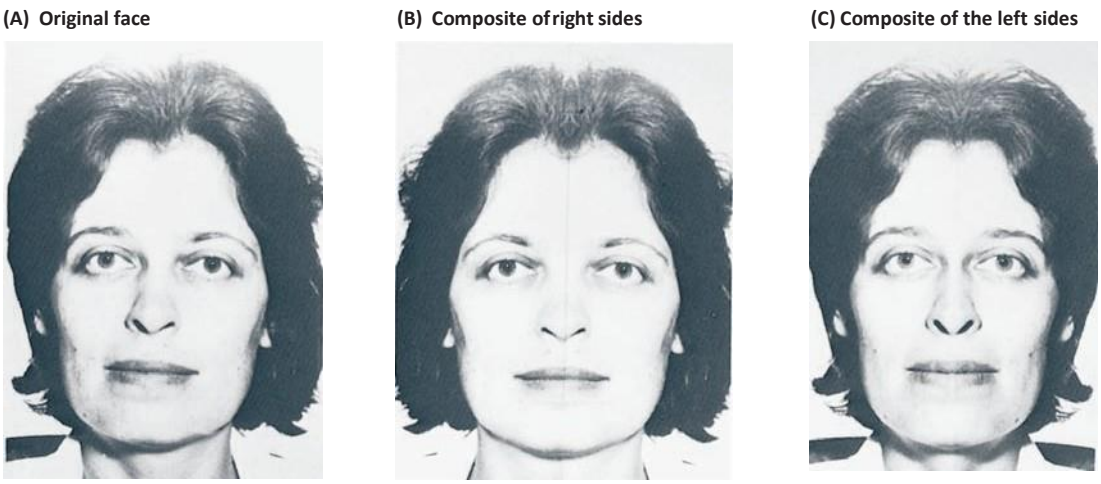


Figure 13.17 The split-faces test. Subjects were asked which of the two pictures, B or C, most closely resembles picture A. Control subjects chose picture C significantly more often than picture B. Picture C corresponds to that part of picture A falling in a subject's left visual field. The woman pictured chose B, the view that she is accustomed to seeing in the mirror. (After Kolb, Milner, and Taylor, 1983.)

Finally, a clear asymmetry exists in the role of the temporal lobes in the analysis of faces. Right temporal lesions have a greater effect on facial processing than do similar left temporal lesions. Even in control subjects, researchers can see an asymmetry in face perception.

We presented subjects with photographs of faces, as illustrated in Figure 13.17. Photographs B and C are composites of the right or the left sides, respectively, of the original face shown in photograph A. Asked to identify which composite most resembled the original, normal subjects consistently matched the left side of photograph A to its composite in photograph C, whether the photographs were presented upright or inverted. Furthermore, patients with either right temporal or right parietal removals failed to consistently match either side of the face in either the upright or the inverted presentation.

The results of this split-faces test not only show an asymmetry in facial processing but also speak to the nature of our perceptions of our own faces. Self-perception provides a unique example of visual perception, because your own image of your face comes largely from looking in a mirror, where the image is reversed, whereas the image that others have of your face comes from direct view. Inspection of Figure 13.17 illustrates the implications of this difference. Photograph A is the image that other people see of this woman and, because there is a left-visual-field bias in our perception, most right-handers choose photograph C as the picture most resembling the original. Consider the choice of the woman herself, however. Her common view of her face (in the mirror) is the reverse of ours, and hence she is more likely to choose (and in fact did choose) composite photograph B as most resembling her own face.

An intriguing consequence of our biased self-facial image is our opinion of personal photographs. Many people complain about not being photogenic, that their photographs are never taken at the correct angle, that their hair wasn't just right, and so on. The problem may be rather different: we are accustomed to seeing ourselves in a mirror image and hence, when we view a photograph, we are biased to look at the side of the face that we do not nor-

mally perceive selectively in the mirror. Indeed, we appear not to see ourselves as others see us. The more asymmetrical the face, the less flattering the person will see his or her image to be.

Visual Imagery

Our ability to conjure up mental images of things that cannot be perceived is central to human thought. Visualization is crucial in problem-solving tasks such as mental arithmetic, map reading, and mechanical reasoning. How crucial can be seen in a patient such as D. F., who was unable to copy drawings or to recognize actual objects but who could nonetheless produce drawings of the same objects from memory (see Figure 13.12).

Behrmann and colleagues described another such patient, C. K. The curious thing about C. K. is that, although he cannot recognize objects, he can imagine them and can draw them in considerable detail from memory. This ability implies some dissociation between the neural system dealing with object perception and that dealing with the generation of images. We can conclude that neural structures mediating the perception and visualization of objects are unlikely to be completely independent, but it is clear that a deficit in object perception cannot be due simply to a loss of mental representations (that is, memory) of objects.

There has been considerable controversy over whether mental rotation of objects might be localized to some region of the right hemisphere. In her review of this literature, Farah concludes that the studies have been "distressingly inconsistent." She proposes that mental rotation probably entails both hemispheres, with some degree of right-hemisphere superiority. (For more detail, see both Farah and Kosslyn.)

Nonetheless, it does seem likely that mental rotation implicates structures related to the dorsal stream. We can imagine that, before a brain could visualize rotating an object, it would first have to have actually rotated it manually. It is a small step to presume that visualizing an object rotating requires the activation of at least part of the motor cortex — the regions needed to actually do it.

In the past two decades, cognitive neuroscientists have conducted a flurry of imaging studies designed to identify the neural events underlying the generation of a mental image. Farah concludes that, although the data are noisy, a reasonably consistent answer is emerging from the results of imaging studies such as the one described in the Snapshot on page 342. Mental imagery appears to be a top-down activation of a subset of the brain's visual areas. In other words, at least some cortical areas are used both for perception and for visualization.

These common areas carry the same representational functions for both purposes, carrying information specifically about color, shape, spatial location, and so on. There is evidence for a distinct mechanism for image generation as well, one separate from the processes needed for perception. Farah notes that the evidence, although mixed, points to a region in the left temporo-occipital region as the key location for this mechanism.

Generating Mental Images

What is the neural basis for visual imagery? It may result from activity in the same visual areas that are active when an image is actually viewed. Another possibility is that some other region of the brain is selectively active when we imagine.

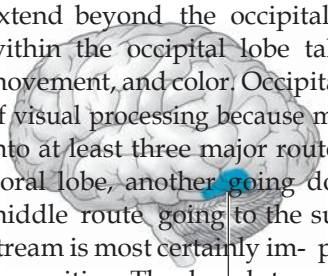
D'Esposito and colleagues addressed this question in an fMRI study by asking subjects to generate mental images from memory, cued by an aurally presented word such as "tree." These cues were common objects rather than abstract representations ("tree" rather than "love," for example). The subjects kept their eyes closed throughout the experiment so that any neural activation could be attributed to imagery rather than to direct activation of the visual pathways. In the baseline condition, the subjects heard abstract words that would not easily allow any image formation, and they were asked simply to listen to the words.

Left temporo-occipital region (fusiform gyrus, area 37)

The results (illustrated here) show that visualizing concrete words increases activation in the left posterior temporo-occipital region, corresponding to the fusiform gyrus (area 37). There was no activation in area V1. The fMRI data are consistent with those of other imaging studies, as well as with a case history of a patient with a left-occipital lobectomy (including area 37) who had a hemianopia in both real and imagined stimuli. The pronounced asymmetry is consistent with Farah's hypothesis that, in most people, the left hemisphere is specialized for image generation.

(M. D'Esposito, J. A. Detre, G. K. Aguirre, M. Stallcup, D. C. Alsop, L. J. Tippet, and M. J. Farah. A functional MRI study of mental image generation. ***Neuropsychologia*** 35:725–730, 1997.)

Summary



The function of the occipital lobe is vision, but visual functions extend beyond the occipital lobe. Separate anatomical regions within the occipital lobe take part in the perception of form, movement, and color. Occipital structures are merely the beginning of visual processing because multiple visual systems can be divided into at least three major routes, one going ventrally into the temporal lobe, another going dorsally into the parietal lobe, and a middle route going to the superior temporal sulcus. The ventral stream is most certainly im- plicated in various aspects of stimulus recognition. The dorsal stream is for the guidance of movement in space. The representation of spatial information re- lies on the recognition of cues within the environment, which would therefore make visuospatial recognition dependent on processing in the ventral stream. An important aspect of the dorsal–ventral distinction in visual processing is that neither route is a single system. Rather, clearly dissociable subsystems take part in various functions. Finally, some occipital regions, especially those ad- joining the temporal cortex, may be functionally asymmetrical. In particular, there appears to be some specialization for word recognition on the left and fa- cial recognition and mental rotation on the right.

Ms. P., a 19-year-old woman, was referred to us by a friend. She was working as a nurse's aide and had found her work so enjoyable that she was considering entering a nursing program. Because she had not completed high school and generally had a poor academic record, she came to us for guidance in deciding whether she could handle such a program. Ms. P. had particular difficulty with language skills, and her reading was so bad that she was unable to pass the written examination for a driver's license. In view of Ms. P.'s interest in furthering her nursing education, we decided to test her reading abilities and to administer a complete neuropsychological battery. The results showed an overall IQ of 85 on the Wechsler Adult Intelligence Scale, but there was a 32-point difference between her verbal IQ of 74 and her performance (nonverbal) IQ of 106. Specific tests of left-hemisphere function confirmed this discrepancy: although her verbal memory, verbal fluency, spelling, reading, and arithmetic scores were extremely low, her spatial skills were good, as were her nonverbal memory and her performance on tests such as the Wisconsin Card-Sorting Test and the Semmes Body-Placing Test. In short, her language skills were those of a 6-year-old, although she had attended school for 11 years, but her other abilities were normal for a person of her age.

In view of her deficient language skills, we concluded that Ms. P. was currently not capable of handling a nursing program. We also felt that she was unlikely to be able to develop the necessary language skills, especially because—as we inadvertently discovered—none of her five brothers and sisters could read either. We explained to Ms. P. that she was by no means retarded but that, just as some people had poor musical ability, she had poor verbal ability. (We were able to arrange an aural administration of the driver's test, which she passed.) Finally, we explained Ms. P.'s problem to her husband, a well-educated man with a master's degree. In the short time that they had been married, he had become totally frustrated with her inability to balance the bank account, read recipes, and so forth, and he was beginning to believe that his wife was either "crazy or retarded." They now had an understanding of the problem, which we hoped would help them work out domestic routines to minimize its effect.

The case of Ms. P. illustrates one type of a *developmental disorder,* a disorder that seems to have its origin in some abnormality in the way in which the brain develops. Hers was a relatively restricted difficulty with language skills that nevertheless must have made school an arduous and frustrating experience and that was clearly continuing to cause problems for her as an adult. In this chapter, we will survey a number of developmental disorders, including disorders of learning, attention, social behavior, and general intellectual functioning.

Learning Disabilities

Large numbers of children enter schools in which they are required to master a core curriculum. Some of them are completely unable to meet any demands of the school system that they enter; some learn, but only with great difficulty; some have to repeat one or more grades; some graduate but fail to master certain subject areas; and some even graduate without mastering basic knowledge in any area. For those who fail, the educational experience often leaves emotional and attitudinal scars that are carried throughout life.

The difficulties that children encounter in school can have any of a number of causes. A child may be disturbed by an unhappy home life, be bored by school, dislike school, dislike a teacher, have no aptitude for school, have low "intelligence," or have a physical handicap or the child may have brain dysfunction or brain damage. Some school systems may be equipped to assess and deal with these kinds of problems. Most have no resources for either assessment or remediation. Even when a school is not equipped to deal with learning problems, neuropsychology now receives enough publicity that, if a child is not learning effectively, the question whether the cause is brain damage or dysfunction or something else will probably arise.

Historical Background

Learning disability is an umbrella term used for a wide variety of school-related problems. Formal definitions of learning disabilities describe these diagnoses as applying to people who have adequate intelligence, opportunity to learn, instruction, and home environment, yet still do not succeed in acquiring certain scholastic abilities. These features are illustrated by the World Federation of Neurology's definition of dyslexia:

> A disorder manifested by difficulty in learning to read despite conventional instruction, adequate intelligence, and sociocultural opportunity. It is dependent upon fundamental cognitive disabilities which are frequently of constitutional origin.

Nevertheless, every key word of this definition and similar ones has been disputed. For example, why should a learning disability be called a disorder? What is meant by conventional instruction? What is meant by adequate intelligence? And so on. To comprehend the difficulty of arriving at satisfactory definitions, knowing some of the history of ideas about learning disabilities can be helpful. Our encapsulation of that history will focus on **dyslexia** (*dys,* "poor function," and *lexia,* "read"), an inability to read properly, because reading is central to many aspects of formal schooling as well as to other aspects of modern life.

Critchley points out that the term *dyslexia* emerged within the context of aphasia, the partial or complete loss of speech, and so it was thought of as being due to damage that occurred at an early age in brain areas responsible for language. In the late 1890s, when James Hinshelwood, a Glasgow eye surgeon, and Pringle Morgan, a Seaford general practitioner, first observed students who could not learn to read, they assumed that their reading failed to develop because the prerequisite brain areas were absent or abnormal. It seemed logical to conclude that **developmental dyslexia**—dyslexia acquired before birth or during early postbirth years—is similar in nature to **acquired dyslexia**—dyslexia due to brain damage after reading has been achieved. Developmental deficits in other spheres, such as mathematics, also would be due to some underlying brain problem.

In the 1920s and 1930s, Samuel T. Orton proposed that dyslexia is due to delayed function, not anatomical absence. Orton, the director of a medical clinic in Iowa, noted that dyslexia was correlated with left-handedness and with tendencies to reverse or invert letters and words when learning to read or write. He termed such dyslexia **strephosymbolia** (from the Greek, meaning "twisted symbols"). Orton thought that the nondominant hemisphere, usually the right hemisphere, which he postulated had a reversed image of things, was excessively dominant or not sufficiently controlled. He suggested that, if an instructor was clever or persevering, education could establish normal dominance of reading in the left hemisphere, and the problem would be resolved.

When sociologists and educational psychologists became interested in learning disabilities, they supposed that environmental explanations, rather than neurological ones, would account for learning impairments. This stance may perhaps have been motivated by the belief, or hope, that environmental causes could be reversed more easily than neurological ones.

The term "learning disability" had its origins in an address given by Samuel A. Kirk in 1963. Kirk argued for better descriptions of children's school problems, but he excluded children with sensory handicaps and mental retardation from the group that he called learning-disabled. The members of his audience who later joined together to form the Association for Children with Learning Disabilities were influenced both by his address and by his definition and further popularized the label that he had coined.

The search for causes of learning disabilities has resulted in a proliferation of terms whose purpose seems to be to dissociate the learning-disabled from the retarded and brain damaged. Fry published a tongue-in-cheek "Do-It-Yourself Terminology Generator," shown in Table 24.1 (from which about 2000 terms can be constructed), to emphasize the overabundance of terms currently flooding the field and the consequent increase in confusion and inaccuracy. Frequently, terms that are widely used take on a pejorative connotation and are then dropped in favor of a new term.

Table 24.1 "Do-it-yourself terminology generator"

Secondary	Nervous	Deficit
Minimal	Brain	Dysfunction
Mild	Cerebral	Damage
Minor	Neurological	Disorder
Chronic	Neurologic	Desynchronization
Diffuse	CNS	Handicap
Specific	Language	Disability
Primary	Reading	Retardation
Developmental	Perceptual	Deficiency
Disorganized	Impulsive	Impairment
Organic	Visual-motor	Pathology
Clumsy	Behavior	Syndrome
Functional	Psychoneurologic	Complex

Directions: Select any word from first column, add any word from second and third columns. If you don't like the result, try again. It will mean about the same thing.
Source: Fry, 1968. Reprinted with the permission of Edward Fry and the International Reading Association.

Incidence of Learning Disabilities

Most estimates of the number of students needing special training to overcome learning disabilities range from 10% to 15% of the school-age population, although only about 2% actually receive special education.

One problem that complicates the calculation of prevalence estimates is that a learning disability is an emerging condition. When children enter the first grade, few of them qualify as being learning-disabled, largely because a popular method of defining a learning disability is to apply a 2-year cutoff criterion: if an person is 2 years behind in academic progress as determined by a standard test, then that person is learning-disabled. When this criterion is used, fewer than 1% of 6-year-olds are disabled, 2% of 7-year-olds are disabled, and so on, until, at age 19, 25% qualify as being disabled. This pattern of emerging incidence develops because the learning-disabled are falling behind at a rate that is proportional to their degree of impairment.

Further complicating the process of calculating and utilizing prevalence rates is the variation in scholastic achievement from one school system to another. Achievement tests are often used to determine grade-equivalent performance, but even nondisabled school populations do not all display equivalent performance. Prevalence rates might be obtained by asking teachers to report the number of children in their class who are receiving special help, but many schools cannot provide such information, because they have no resources for special education.

Types of Learning Disabilities

The classification and incidence of learning disabilities correspond to the emphasis placed on self-control and on certain academic specialties in the public school system. Good behavior, reading, arithmetic, and spelling are emphasized, and learning-disability classification reflects this focus. Although art, music, and physical education are taught in many schools, referrals for failure in these areas are uncommon. If art, rather than reading, were the core subject in the early years of school, we suspect that current catalogues of types of disabilities would be different. Nevertheless, there *are* disabilities that interfere with the acquisition of reading, spatial orientation, mathematics, and social skills.

In summarizing the syndromes seen in a 2-year period at a clinic specializing in learning disabilities, Denckla reports that, of 484 children aged 6 to 16 years, 76% were classified as primarily dyslexic, with or without some associated problems, and 18% were classified as hyperactive. Thus, reading and behavior problems were the most common reasons for referral. The American Psychiatric Association's *Diagnostic and Statistical Manual of Mental Disorders* (DSM-IV) recognizes a number of categories of disorders arising in childhood, including mental retardation and mental disorders; disorders in reading, arithmetic, and motor activity; and certain mixed classifications. The U.S. Department of Health, Education, and Welfare lists 10 characteristics most often cited by various authors as being associated with a learning disability: (1) hyperactivity; (2) perceptual–motor impairments; (3) emotional lability; (4) general coordination deficits; (5) disorders of attention (short attention span, distractibility, perseveration); (6) impulsivity; (7) disorders of memory and thinking; (8) specific learning disabilities, including, especially,

those of reading (dyslexia), arithmetic, writing, and spelling; (9) disorders of speech and hearing; and (10) equivocal neurological signs and irregular EEG. Not all learning-disabled children exhibit all these symptoms. For example, for every learning-disabled child with coordination problems, there is a learning-disabled child whose coordination is better than normal.

Reading Disabilities

Reading requires letter-identification skills, phonological skills (converting letters into sounds by using certain rules), grapheme association skills (using the visual gestalt of a word to access a previously learned sound), sequencing skills (in which a number of sounds are analyzed and combined in sequence), and short-term-memory skills (to retain pieces of information as they are sequentially extracted from written material). Acquired information also is important, including knowledge of words in the form of a **lexicon.** A dictionary-like store of words in the brain, a lexicon contains their meanings, knowledge of the way in which they can be combined, and information about the ideas with which they can be associated. Thus, reading is a multiprocess and multistage behavior. As such, one would expect that it could be disrupted in many different ways. In the following sections, we will describe (1) types of reading, (2) the role of sensory detection in reading, (3) other deficits that correlate with deficits in reading, and (4) the role of neuropsychological evaluation in reading.

Types of Reading

Reading can be accomplished in either of two ways. Consider the following example. Imagine that you are reading a novel about a man named Fzylx from Worcester. Let us assume that you have never encountered either the man's name or his hometown before. When you read the names, you presumably attempt to sound out the sequences of letters to arrive at a satisfactory pronunciation of the words. Suppose that the next day you overhear two people discussing a novel about a man named Fzylx from Worcester. At first, you might be struck by the similarity between the plot of your book and theirs, until you confront them and find out they are actually discussing Felix from Wooster! What has happened? When you read Fzylx and Worcester, you encountered two common problems in reading English. First, you had to read a name that is not English and so you had no rules by which to read it. Even so, you may have felt comfortable with your pronunciation, inasmuch as it allowed you to read the name. Second, you encountered an irregular word; that is, a word that is not pronounced the way that it is spelled. The only way to pronounce this type of word correctly is to memorize it. This type of reading is called **graphemic reading** (or sometimes *lexical reading*). It can be used for regular words as well, but it *must* be used for irregular words or words that are not found in English. It is also the way Arabic numerals (such as 4) and international symbolic road and direction signs must be read. The other way to read is to do what you tried to do with Fzylx and Worcester. You simply convert a letter or group of letters into sounds (phonemes) that will provide the clue to

the meaning of the words. Stated differently, the sounds that you get by analyzing letter groups will lead you to a pronunciation, and you will be able to access your memory, or lexicon, for the meaning and connection of the word. This is known as **phonological reading.**

Bradley and Bryant suggest that the phonological procedure is used by beginning readers but that, as reading skill is attained, the graphemic procedure becomes more important. They suggest that the transition takes place between 6 and 10 years of age. Thus, normal reading initially requires phonological skills and later becomes dependent on grapheme skills. This progression may explain why many people have difficulty in finding typographical errors when they proofread. Rather than reading phonologically, they read graphemically and, practiced at graphemic reading, they need to read only part of a word before recognizing its meaning and shifting attention to the next word. If the spelling error is not within the part of the word actually read, it will not be noticed. As we shall see shortly, there is evidence that phonological reading may be a function of the left hemisphere and grapheme reading a function of the right. In some school systems, instruction begins with grapheme reading, skipping the phonological stage, a procedure that has ignited considerable controversy regarding the appropriate way to teach reading.

Given the differences between these two reading processes, we should find at least two different kinds of reading impairments, which, moreover, could arise at different ages. A child who is incompetent in the phonological procedure will have difficulty in the early stages of reading. A child who is competent in the phonological procedure but incompetent in the grapheme procedure will have difficulty later on. Frith demonstrated just such types of age-related disabilities among poor readers. It may also follow that a child who is impaired at the first type will be hampered in making the transition to the second type.

We should note here that these types of impairments would not by any means exhaust the classifications of poor readers. People with a poor short-term auditory memory may not make proper sense of written material, because they quickly forget the words and phrases as they proceed. This type of disability may be particularly obvious at older ages, when reading material becomes more complex. People with poor long-term memory may not understand the sense of words despite good decoding skills, simply because they do not have much information about the meaning of the words. This is similar to the situation of a person who speaks only English trying to read Italian. The person would be able to sound out the words by using general phonetic rules but would not understand what he or she was reading, not knowing what the words meant. In fact, people who are demented are often just like this. They can read, but they understand nothing.

The Role of Sensory Detection in Reading

Impairments in language and reading may stem from difficulties that children have with sounds. Bradley and Bryant tested the sound-categorization ability of children who had not yet started to read. The tests consisted of giving a child three or four words and asking him or her to pick out the word that does not have a sound (phoneme) in common with the others. For example, in the

series "hill, pig, pin," "hill" would be the correct choice; in the series "cot, pot, hat," "hat" would be the correct choice, and in the series "pin, bun, gun," "pin" would be the correct choice. Bradley and Bryant found that, when the same children were older and had started to learn to read, those who were initially weak at sound categorization were the ones who later fell behind in reading and spelling. They argue that the initial insensitivity to rhyme and alliteration causes subsequent reading impairment, because, if the children who were initially impaired were given special training in rhyme and alliteration, their reading was far less impaired after training in reading began. In short, according to Bradley and Bryant, at least one cause of reading deficiency is a basic deficiency in phonological, or sound, awareness.

It is possible that children who are at risk for learning disabilities could be detected still earlier. Frith suggested that the babbling of babies may give clues about those who are at risk. Werker and Tees reported that young infants can detect differences between all known speech sounds, but, as they acquire language, their detection abilities become restricted to only those sounds used in their own language, to which they are exposed daily. Impairments in this refining of their detection abilities could lead to language difficulties.

Tallal and her coworkers examined the sensory detection abilities of children with learning disabilities and found that they are impaired in detecting sensory events that take place in rapid succession. If two sensory stimuli, such as two tones, are presented in succession very quickly, they will be heard as one tone. If the interval between the tones is gradually increased, a point will be reached at which they are heard as two tones. For most people, a separation of tens of milliseconds, from about 10 to 40 ms, is required before the two tones are discriminated. For people with language impairments, a much greater separation is needed (Figure 24.1). The relevance of this finding to language impairments is that stop consonants ("ba," "da," "ga," "pa," and "ta") contain a transition period in which the sounds (called formants) change very rapidly, usually within 40 ms. When stop consonants are used as stimuli, language-impaired people have difficulty in differentiating one such consonant from another, whereas they have no difficulty in detecting vowels. They also have no difficulty in detecting stop consonants if the transition period is lengthened. When this difficulty is detected in infants, it is predictive of later language impairment.

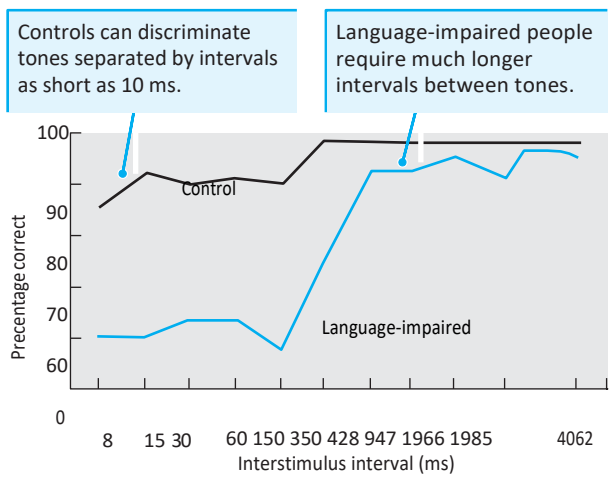


Figure 24.1 Percentage of trials in which controls and language-impaired subjects discriminate two tones separated by different interstimulus intervals. (After Tallal et al., 1993.)

Tallal and coworkers suggest that the left hemisphere is specialized for making these rapid sensory discriminations and movements. Thus, it has the requisite capacity to become dominant for language. It follows that impairments of the left hemisphere will impair discrimination abilities that are important for both the acquisition and the production of language.

This idea points to possible strategies for remediation. Tallal and coworkers suggest that remediation of language-related disorders should focus on training in discrimination rather than on more generalized training in language-related skills. For example, they reason that, if a child has difficulty in discriminating "da" from "de" when the sounds are presented

at a normal rate, then slowing the presentation down so that the duration of the sounds is dragged out would make discrimination easier. They constructed computer games in which the subjects, rewarded each time that they made a correct discrimination (either between verbal or between nonverbal stimuli), gradually worked their way from simple to more-complex discrimination problems. They also designed computerized listening exercises that taught phonological discrimination through the use of acoustically modified speech. The computer-based exercises measured each subject's initial performance level and then led the subject through extensive daily training for a number of weeks, after which the subject's performance level was found to be closer to normal.

Training in sound-processing rates resulted in improvement in temporal integration on the discrimination tasks. The training also resulted in improvement in language comprehension abilities as assessed with the use of independent tests. In follow-up tests given approximately 6 months after training, the improvements were found to have endured. A group of subjects that received equivalent training in which natural rather than modified sounds were used showed no equivalent improvement (see the Snapshot on page 650).

The efficacy of this program—called Fast ForWord—has been tested in a large-scale trial conducted in 35 educational settings in the United States and Canada. A total of 500 children were trained for about 2 hours a day for as long as 2 months. The results indicated that the children improved in discrimination ability as a result of the training; they also improved in language skills as measured by independent tests. These findings suggest that improvement in sound discrimination can lead to improvement in language abilities.

Analysis of Correlated Deficits

It would be helpful if reading deficits displayed themselves in a straightforward manner, but unfortunately, they do not. People with dyslexia exhibit a wide range of different symptom clusters, as well as considerable individual variation. We cannot consider each of the many symptoms here, but they include deficits in attention, eye movement, development, memory, coordination, spatial abilities, movement sequencing, map reading, and visuospatial processing. The reason that language disorders are associated with so many kinds of symptoms is that language has a high-level role in managing our mental processes and thus affects many different kinds of behavior.

Impairments in separating stimuli have been found in all sensory modalities in dyslexic persons, as have impairments in the production of movements. Thus, language-impaired persons may require longer intervals between stimuli before they can detect two separate lights or two separate touches, and they may be impaired in producing rapid movements. They may also be impaired in sound-frequency discriminations and in detecting a target sound obscured by background noise. Hari and Renvall, in reviewing the many sensory and motor deficits associated with dyslexia, suggest that the central problem could be "sluggish attention shifting." In other words, when the attention of these dyslexic subjects is engaged, it cannot easily be disengaged, and vice versa. They suggest that the problem arises in the associational areas of the parietal lobe, which receive input from all sensory systems and then initiate movements. A

SNAPSHOT

Imaging Sound Perception in Normal and Dyslexic Subjects

Developmental dyslexia—difficulty in learning to read—affects between 5% and 20% of the population, yet its neural basis is not known. One hypothesis is that dyslexics are impaired in an aspect of phonological processing known as phonemic awareness, defined as the ability to consciously ***decompose*** words into their constituent speech sounds. Phonemic awareness develops as children learn to read, specifically as they learn to map written words onto existing representations of spoken language within their brains. The results of some research suggest that phonological processing deficits are indications of a more fundamental deficit in processing rapidly changing acoustical signals, an ability that is necessary for language comprehension. This hypothesis, known as the ***rapid processing hypothesis***, led to the proposal that subjects given training in the discrimination of rapidly changing acoustical signals would improve in their ability to discriminate between the signals and in their comprehension of auditory language.

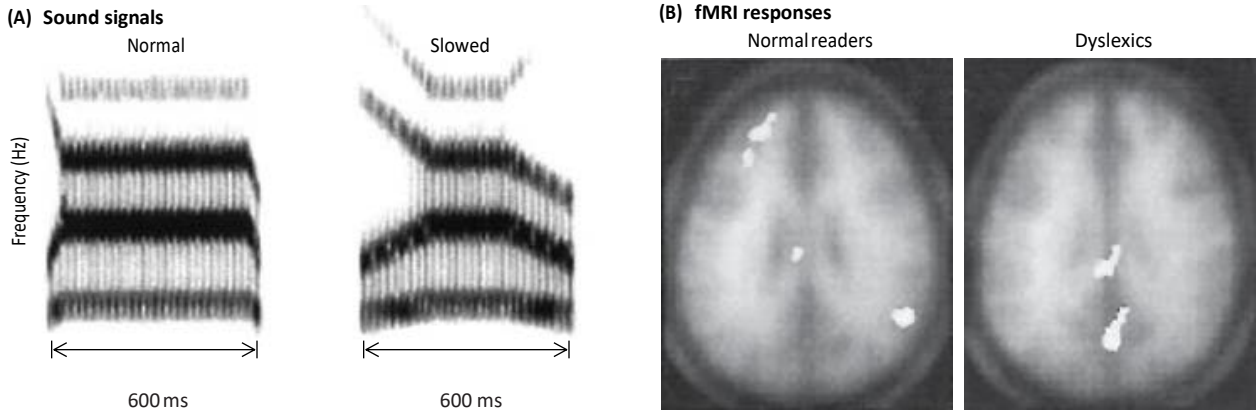
Temple and colleagues tested 8 adult subjects with a history of developmental dyslexia and 10 matched control subjects on their discrimination of rapid acoustical signals, while obtaining functional magnetic resonance images of the subjects' brain activity. The images revealed a specific disruption of the neural responses to transient, rapidly changing acoustical stimuli in the adults with developmental dyslexia. The largest activation in the control subjects was in the left prefrontal region, between the middle and superior frontal gyri in Brodmann's areas 46-10-9. Analysis of the dyslexic readers revealed no increase in left frontal response to the rapid relative to the slow stimuli. The illustration shows examples of rapid and slowed nonspeech acoustical signals (A) and examples of normal and dyslexic subjects' fMRI responses to rapid auditory stimuli (B). Note the greater activation of the left prefrontal cortex in normal readers.

Three of the dyslexic subjects then underwent a training program designed to improve rapid processing, after which they were again examined with the use of fMRI. The training consisted of computer exercises—100 minutes a day, 5 days a week, for 33 days—designed to improve rapid processing of linguistic and nonlinguistic stimuli. After training, two of the subjects improved on tests of rapid auditory processing and auditory language comprehension, and their fMRIs showed significantly increased activity in the left prefrontal cortex. The third subject showed no in-

problem there, they believe, could lead to an inability to switch attention that would affect many sensory domains as well as the production of movements.

It is important to emphasize that none of the theories concerning the underlying causes of reading impairments is uncontroversial. An understanding of learning disorders is fraught with complexity, as can be illustrated with a brief account of the magnocellular theory (a complete discussion of which would be far beyond the scope of this book). This theory states that reading disorders stem from problems in the magnocellular part of the visual system,

which in normal function responds to rapidly changing stimuli. If a child's detection of visual motion is disturbed because of a dysfunction there, the child may have difficulty in reading because the words on the page appear to jump around. The theory also suggests that, if reading is done through a color filter or with only one eye, the perception of word movement can be reduced and reading can be improved. Skottum, in reviewing both the theory and its sup-



crease in left frontal activity and no behavioral improvement. These results not only suggest a role for the left prefrontal cortex in processing nonlinguistic rapidly changing acoustical stimuli but also indicate that fMRI can be used for the diagnosis and treatment of reading disorders.

(E. Temple. Brain mechanisms in normal and dyslexic readers. ***Current Opinion in Neurobiology*** 12:178–183, 2002.)

Analyzing sound perception in normal and dyslexic readers. (A) Rapid and slowed nonspeech acoustic signals used as test stimuli and in training. (B) fMRI responses to rapid auditory stimuli in normal readers and dyslexic subjects. (From E. Temple, R. A. Poldrack, A. Protopapas, S. Nagarajan, T. Salz, P. Tallal, M. M. Merzenich, and J. D. Gabrieli. Disruption of the neural response to rapid acoustic stimuli in dyslexia: Evidence from functional MRI. *Proceedings of the National Academy of Sciences of the United States of America* 97:13907-13912, 2000.)

port, finds that, apart from problems with the theory itself, of 22 studies examining the theory, 4 were in agreement with it, 11 disagreed with it, and 7 were consistent with an alternative explanation. The mixed nature of these results stems from the complexities of the theory, inconsistencies in the choice and diagnosis of subjects, and methodological differences in the various lines of research used to test the theory.

Neuropsychological Evaluation

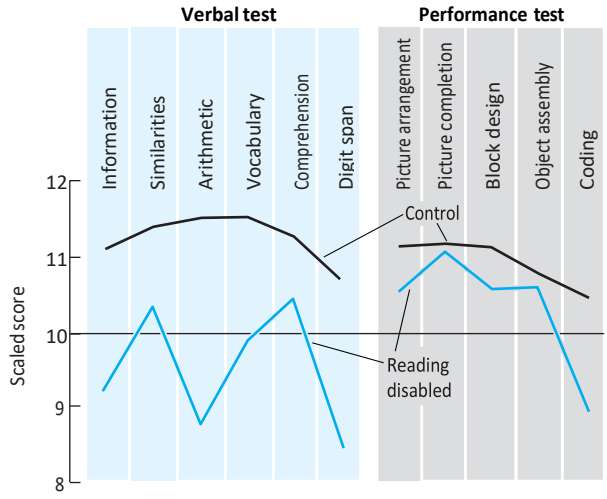
Neuropsychological approaches to assessing dyslexia generally rest on the following assumptions: (1) the disability may affect only one or a few spheres of endeavor; (2) a specific skill or lack of that skill can be detected through a neuropsychological testing procedure; (3) if one method or strategy of instruction

is unsuccessful, another might be more successful; and (4) the neuropsychological

test results should suggest a possible strategy for remediation of the learning disability. Although none of these assumptions has yet received anything like adequate scientific support, the neuropsychological testing strategy does provide a comprehensive evaluation of a person that is useful for counseling, as illustrated in Ms. P.'s case at the beginning of this chapter.

Neuropsychological testing assesses performance on a wide range of tasks, providing feedback on all areas of brain function. Most learning-disabled children are not dyslexic or dyscalculic (can't do math) alone but have a number of associated symptoms of which teachers and parents are usually unaware. The discovery of these associated deficits often helps the adults in the child's life to understand the difficulties with which the child is struggling. Neuropsychological tests can also help distinguish between children who have central reading impairments and those whose problems have emotional or social causes.

Many studies have focused on the IQ test results of learning-disabled children. These analyses attempt to correlate learning impairments with performance on the subtests of the Wechsler Intelligence Scale for Children. Rugel compiled the results from studies in which, collectively, a total of 1521 reading-disabled children and 554 control children were tested and compared. Figure 24.2 presents a graphic summary of these results. The dyslexic group displays low scores on four subtests: arithmetic, coding, information, and digit span. This profile, typical of many such studies, is referred to as the ACID profile. Dyslexic children characteristically have an overall IQ score that averages about 7 points lower than the same score attained by control children, but their mean IQ is roughly 100. We (Whishaw and Kolb) report that children above the age of 8 show the ACID profile, whereas those younger than 8 may not show a deficit in the information or arithmetic subscales. This finding suggests that ACID deficits in older children and adults are secondary to the underlying impairment that produces dyslexia. Although the deficits in digit span and coding are commonly seen with dyslexia, there is no agreement that they are necessarily related to a disability in reading.



	Reading-disabled	Control
Verbal IQ	98	108
Performance IQ	102	106
Full-scale IQ	100	107
Number of subjects	1521	554

Figure 24.2 Intelligence test profiles of developmentally dyslexic subjects and controls. Note the low scores on arithmetic, coding, information, and digit span, referred to as the ACID profile. (From Rugel, 1974; after Whishaw and Kolb, 1984.)

Many researchers have commented on the large differences between verbal IQ and performance IQ in dyslexic persons. Some experts believe that two types of dyslexics can be identified on the basis of these scores. Generally speaking, however, a dyslexic child's subscores will vary greatly (the child will score in the high range on some subtests, in the low range on others, and in the average range on still others). Nevertheless, for an experienced counselor, the pattern displayed by any child may be meaningful.

In comparing the performance of a dyslexic group with that of the control group on other sections of their composite test battery, we (Whishaw and Kolb) found that the tests did discriminate between the two groups but that their doing so depended in part

on the person's age. This age dependence was particularly evident in three tests. In the test of left–right differentiation (Figure 24.3), neither dyslexic children nor age-matched controls could score above chance if they were younger than 8 years old. After the age of 8, the control children performed well, whereas the dyslexic children continued to perform at chance. A different kind of emerging difference was found on tests of word fluency (for example, "Give as many words beginning with the letter 'S' as you can"), in which dyslexic and control scores were similar in children younger than 8 years but diverged increasingly in older age groups, suggesting that the fluency performance

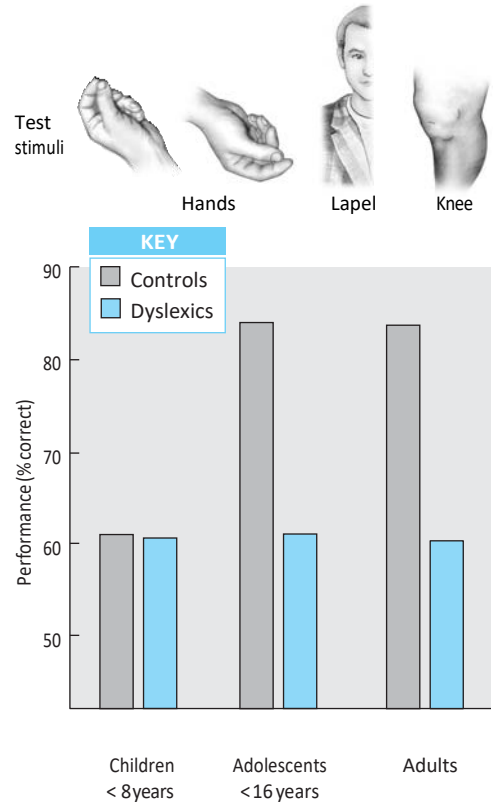


Figure 24.3 Performance on Semmes's left-right discrimination test by control subjects and dyslexic subjects. Stars signify significant group differences. (After Whishaw and Kolb, 1984.)

in the control group improved with age, whereas the dyslexic group remained almost static. A third pattern was obtained on the Semmes Body-Placing Test. Here (a test of left–right discrimination) significant group differences emerged only in adults, and these differences seemed to depend on the fact that adult control subjects displayed virtually perfect performance on the tests. These observations suggest to us that, although the tests can be applied with some success to children, they must be interpreted with caution in regard to younger children, and retesting at different ages is worthwhile.

Nonlanguage Learning Disabilities

In the six nonlanguage learning disabilities described in this section—that is, (1) hyperactivity, (2) cerebral palsy, (3) hydrocephalus, (4) autism, (5) fragile-X syndrome, and (6) fetal alcohol syndrome—children have difficulty in comprehending aspects of their environment, in pretending and anticipating, in interpreting the facial and emotional gestures of others, and in performing skilled movements.

Hyperactivity

Hyperactive child syndrome is distinguished from other types of learning disabilities in that an affected child is a behavioral problem in school, and all aspects of school performance are usually disrupted. This syndrome is sometimes called attention deficit disorder (ADD) or attention deficit

hyperactivity disorder (ADHD). Hyperactive children may have specific learning disabilities in addition to the hyperactivity, and these disabilities possibly contribute to it. A number of diagnostic labels have been given to this disorder, including *minimal brain dysfunction, hyperkinetic child syndrome,* and *hyperkinetic impulsive disorder.* The DSM-IV lists the following diagnostic criteria:

1. *Excessive general hyperactivity or motor restlessness for the child's age.* In preschool and early school years, there may be incessant haphazard, impulsive running, climbing, or crawling. During middle childhood or adolescence, marked inability to sit still, up and down activity, and fidgeting are characteristic. The activity differs from the norms for the age both in quality and in quantity.
2. *Difficulty in sustaining attention, such as inability to complete tasks initiated or a disorganized approach to tasks.* The child frequently "forgets" demands made or tasks assigned and shows poor attention in unstructured situations or when demands are made for independent, unsupervised performance.
3. *Impulsive behavior.*
4. *Duration of at least 1 year.*

In infancy, hyperactive children are thought to exhibit poor and irregular sleep, colic, and feeding problems and to not like being cuddled or held still for long. Later, they are described as learning to run rather than to walk and as being driven to handle and play with everything. By the time they reach kindergarten, they are demanding, do not listen, and do not play well with other children. People outside the home may begin to reject a hyperactive child because of his or her behavior. By the time the child enters school, his or her high level of activity, low tolerance for frustration, poor concentration, and poor self-esteem lead to a referral for assessment. By adolescence, many of these children are failing in school, and from 25% to 50% of them have begun to encounter problems with the law. Their behavior remains restless, they withdraw from school, and they fail to develop social relations and maintain steady employment.

The hyperactive syndrome is described by Weiss and Hechtman as the most common behavioral disturbance among children. Estimates of its incidence vary because of different definitions and cultural differences in tolerance of hyperactive behavior. Estimates of the ratio of boys to girls range from 5:1 to 9:1. In the Isle of Wight study conducted by Rutter, an overall population incidence of 1 in 1000 was reported, but, in North America, where tolerance of hyperactive behavior seems lower, estimates of incidence are as high as 6 in 100. It does seem that North American parents' and teachers' estimates of what constitutes normal behavior may be unrealistic, because Weiss and Hechtman note that, in surveys of parents and teachers in North America, as many as 50% of children are reported as being hyperactive.

The suggested causes of hyperactivity include brain damage, encephalitis, genetics, food allergies, high lead concentrations, and various home and school environments. A single cause is unlikely to be responsible for all cases. Therapy includes counseling for the child and parents and careful structuring of the

home and school environments. Beginning in the 1960s and continuing to the present, treatment with amphetamine-like stimulant drugs such as Ritalin has been popular, although the effectiveness of Ritalin or other drug treatment as a long-term solution is doubtful. When Ritalin *is* effective, it may be because, as a stimulant, it allows the person to concentrate on the task at hand. The drug may also have a general sedating effect on children.

Cerebral Palsy

Cerebral palsy is a disorder primarily of motor function caused by brain trauma in the course of fetal development or birth. Any simple definition is difficult, however, because (1) the motor symptoms take many forms, (2) there can be various kinds of accompanying cognitive impairments, and (3) the causes are diverse. In consequence, cerebral palsy cannot be accurately called a disease, a syndrome, or even a condition; it will take a different form in each person, depending on the nature of the brain damage. The term "cerebral palsy" is therefore most useful in an administrative sense, as a category of persons who are handicapped in many different ways by motor disorders due to nonprogressive brain abnormalities. Because brain damage is the underlying cause, cerebral palsy is not curable, but it often is amenable to therapy and training.

Cerebral palsy was first described in the medical literature in 1853 by London physician William Little. He recognized that the motor abnormalities of some babies are a result of abnormal parturition, difficult labor, premature birth, or asphyxia. He also recognized the permanence of the disabilities and their associated intellectual impairments; effects on personality (such as irritability and temper tantrums); and epilepsy. More important, he pointed out that the problems could be severely aggravated by subsequent improper training and education.

The incidence of cerebral palsy is estimated at about 6 per 1000 births. The numbers of males and females afflicted are about equal. Estimates of the degree of impairment suggest that about 10% of afflicted persons require no special services, 65% need services on an occasional basis, and about 25% need special schooling or custodial care. When cases of cerebral palsy are categorized by motor symptoms, about 50% of persons with the disorder are spastic (their limbs resist being moved), about 25% are athetoid (they make slow involuntary movements), about 10% are afflicted with rigidity (muscles around joints are stiff), and about 10% are ataxic (have difficulty making voluntary movements). As noted earlier, cerebral palsy has many causes, the most frequent of which are listed in Table 24.2. Nearly 50% of all cases are due to birth injury or injury suffered during development, 9% are secondary to convulsions, and 8% are due to prematurity. Smaller numbers result from other diverse causes. Incidence is also related to the mother's ability to carry a baby to term and to factors such as her body size, health habits, and weight gain during pregnancy.

Lesions of the corticospinal tracts, basal ganglia, brainstem, and cerebellum are presumed to be responsible for the disorders, but it has been difficult to establish clear-cut relations between lesions and clinical findings.

Table 24.2 Potential causes of cerebral palsy

Hereditary

Static—familial athetosis, familial paraplegia, familial tremor

Progressive—demyelinating diseases of viral or undetermined origin (chromosomal breakages are rare in cerebral palsy, as are disorders of metabolism)

Congenital (acquired in utero)

Infectious rubella, toxoplasmosis, cytomegalic inclusions, herpes simplex, and other viral or infectious agents

Maternal anoxia, carbon monoxide poisoning, strangulation, anemia, hypotension associated with spinal anesthesia, placental infarcts, placenta abruptio

Prenatal cerebral hemorrhage, maternal toxemia, direct trauma, maternal bleeding, diathesis

Prenatal anoxia, twisting or kinking of the cord

Miscellaneous toxins, drugs

Perinatal (obstetrical)

Mechanical anoxia—respiratory obstruction, narcotism due to oversedation with drugs, placenta previa or abruptio, hypotension associated with spinal anesthesia, breech delivery with delay of the after-coming head

Trauma—hemorrhage associated with dystocia, disproportions and malpositions of labor, sudden pressure changes, precipitate delivery, caesarean delivery

Complications of birth—"small for date" babies, prematurity, immaturity, dysmaturity, postmaturity, hyperbilirubinemia and isoimmunization factors (kernicterus due to Rh factor, ABO incompatibility), hemolytic disorders, "respiratory distress" disorders, syphilis, meningitis, and other infections, drug addiction reactions, hypoglycemic reactions, hypocalcemic reactions

Postnatal—Infancy

Trauma—subdural hematoma, skull fracture, cerebral contusion

Infectious—meningitis, encephalitis, brain abscess

Vascular accidents—congenital cerebral aneurism, thrombosis, embolism, hypertensive encephalopathy, sudden pressure changes

Toxins—lead, arsenic, coal-tar derivatives

Anoxia—carbon monoxide poisoning, strangulation, high-altitude and deep-pressure anoxia, hypoglycemia

Neoplastic and late neurodevelopmental defects—tumor, cyst, progressive hydrocephalus

Source: E. Denhoff, Medical aspects. In W. M. Cruickshank, Ed. *Cerebral Palsy*. Syracuse, NY: Syracuse University Press, 1976, p. 33. Reprinted with permission.

Hydrocephalus

Characterized by an increase in the volume of the cerebrospinal fluid (CSF), **hydrocephalus** can be caused in two ways. In one way, more likely to develop in adults, enlarged ventricles can be a secondary result of shrinkage or atrophy of surrounding brain tissue. In the second, more typical cause of hydrocephalus — and more typical in infants — obstruction of the flow of CSF results in a buildup of pressure in one or more ventricles that eventually causes their expansion. It is not certain that a simple overproduction of CSF is ever a cause of hydrocephalus.

Figure 24.4 is a drawing made from a cast of the lateral ventricles in a normal brain. In a living brain, the ventricles are filled with CSF. The usual amount in an adult is only about 130 cm^3, of which about one-third is in the spinal cord's great lumbar cistern. As described in Chapter 3, CSF is made by the

choroid plexus in the ventricles, most of it in the lateral ventricles. From there it flows through the interventricular foramina (windows) of Monro into the third ventricle, through the cerebral aqueduct, and then into the fourth ventricle. It finally escapes through three little holes in the roof of the fourth ventricle. These holes are the two laterally located foramina of Luschka and the medial foramen of Magendie. (The mnemonic is: lateral, Luschka; medial, Magendie.) The fluid then enters the subarachnoid space—the space beneath the arachnoid covering of the brain and spinal cord. It is absorbed into the veins and carried away by the bloodstream.

The circulation in the ventricles can be blocked at either of the interventricular foramina, causing an increase in pressure followed by expansion of either lateral ventricle. It can also be blocked at the level of the cerebral aqueduct (causing hydrocephalus of the first three ventricles) or by closure of the foramina in the roof of the fourth ventricle (producing hydrocephalus of the entire ventricular system). Any sudden obstruction of CSF flow will cause a rapid rise in intracranial pressure, ventricular dilation, and finally coma. A gradual obstruction, such as by a tumor, causes a less-rapid increase in pressure and consequent dilation, and the symptoms may include the gradual appearance of visual disturbances, palsies, dementia, and so on.

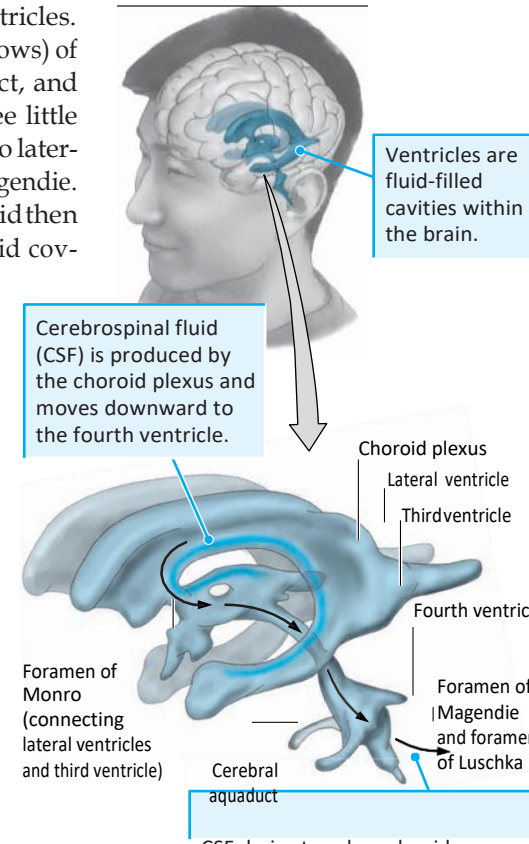


Figure 24.4 Drawing of a cast of the ventricular system of the brain as seen from the side. The arrows indicate the direction of flow of cerebrospinal fluid. A blockage of the flow in the narrower parts of the ventricles (for example, the cerebral aqueduct) can cause the symptoms of hydrocephalus.

Infant hydrocephalus, characterized by a conspicuous enlargement of the head, usually develops during the first few months of life. As many as 27 of 100,000 newborn babies may suffer from it. In about 14% of these cases, a malformation impedes CSF circulation; most other cases are produced by inflammation or trauma, although about 4% are due to tumors. As the ventricles distend, they push the cerebral hemispheres into a balloon shape. Because the skull bones of an infant are not yet fused, continued pressure causes the expansion of the head in all directions. If expansion damages the cortex, intelligence may be impaired and dementia may result; but, if the cortex is not damaged, intelligence may be unimpaired even after the cortex has been stretched into a sheet of tissue less than a centimeter thick. Hydrocephalus can be treated with some success by the insertion into one lateral ventricle of a valve and a tube that passes into a jugular vein to drain into the cardiac atrium. Untreated, it often causes death or severe mental or motor disabilities.

Autism

The term **autism** was first used by Kanner and Asperger in the 1940s to describe individual children without obvious signs of focal cerebral disease and with the symptoms of severely impaired social interaction, a bizarre and narrow range of interests, language and communication abnormalities, and, in some cases, preserved intellect. In summary, children with autism have a hard time communicating with others and functioning in the world outside the home. Autism is estimated to affect as many as 1 in 500 children, is four times as prevalent in boys as in girls, and has no known racial, ethnic, or social boundaries.

Many autistic infants are odd from birth, avoiding physical contact with caregivers by arching their backs or becoming limp when held. Approximately one-third of autistic children develop normally until between 1 and 3 years of age, when the autistic symptoms begin to emerge. Common characteristics are a failure to interact socially and an insistence on sameness. One possible reason for the latter may be an inability to understand and cope with novel situations. Autistic persons may exhibit repeated body movements (hand flapping, rocking), have unusual responses to people or unusual attachments to objects, and resist any changes in routine. In some cases, aggressive or self-injurious behavior or both behaviors are seen as well. Some autistic people are severely impaired, others can function on their own, and still others have exceptional abilities in some areas, such as music, art, or mathematics.

Autism has no known specific cause. There is some indication of a genetic influence, however: autism is more likely to develop in two identical twins than in two fraternal twins. There is also evidence that a virus can cause autism: women have an increased risk of having an autistic child after exposure to rubella in the first trimester of pregnancy. There is some suspicion that autism can be caused by industrial toxins, but the evidence for this cause is uncertain. In addition, the results of research reviewed by Bauman and Kempor suggest that brain abnormalities of various types—especially in the temporal lobes and cerebellum—might correlate with the degree of impairment displayed by an autistic person. In that case, impairments in explicit memories (memories for daily events) might be related to the temporal lobe abnormalities, whereas impairments in implicit memory (skills and conditioned responses) might be related to the cerebellar abnormalities. Evidence that the cerebellum controls conditioned responses suggests that the desire for sameness and the avoidance of novelty also may be related to cerebellar abnormality. One feature of conditional learning is habituation, or learning to ignore irrelevant or repeated stimuli. In the absence of an ability to habituate to ongoing events, a person may find such stimuli especially noxious and so avoid them in favor of maintaining sameness. This theory could explain why autistic people report that the sound of traffic, to which most people quickly habituate, remains for them frighteningly loud.

Roder suggests that one cause of autism may be an abnormality in the expression of genes that play a central role in the development of the brainstem. She finds that an area of the brainstem in the caudal part of the pons is small in autistic subjects and that several nuclei in this area, including the facial nucleus, which controls facial musculature, are small or missing (Figure 24.5). In addition, many autistic children have subtle facial abnormalities that may be due to abnormalities of the facial nerve. Perhaps mutation of the *HOSA1* gene, which plays a role in the development of the brainstem, or interference in the expression of this gene is responsible for many cases of autism.

A less-severe condition of withdrawal in children is referred to as **Asperger's syndrome.** These children, although withdrawn, exhibit early speech and good grammar, but they also exhibit narrow repetitive play, poor peer relations, and a need for routine and sameness. They, too, may excel in some aspect of behavior, such as reading, calculations, music, or art. **Hyperlexia** is a term describing unusual reading ability in otherwise cognitively impaired persons, such as children with Asperger's syndrome. It is marked by a precocious development of reading abilities between the ages of 3 and 5 years. Very often the children teach them-

selves to read. In Asperger's syndrome, hyperlexia is often accompanied by exceptional memory abilities, such as an unusual ability to remember words, television shows, names of streets, the weather, birthdays, and so forth. Reading may not be completely fluid, because there are often articulatory defects and prosodic abnormalities of intonation and rate of speech. Generally, comprehension of reading is impaired, and the children show emotional withdrawal, occasional echolalia (repeating words that they hear), and autistic symptoms.

Related to Asperger's syndrome is the **savant syndrome,** or *idiot savant syndrome,* first described by John Langdon Down in 1887. Since then, several hundred cases have been reported in the literature. The affected persons are remarkably similar in that they have a narrow range of special abilities and a common symptomatic triad of retardation, blindness, and musical genius. *Idiot savant* was coined by combining the word *idiot,* at one time an accepted name for a subcategory of mental retardation, with *savant,* which means a knowledgeable person. The term has endured despite its now pejorative connotation.

Savants are characterized by mental handicaps, resulting from a developmental disability or mental illness, combined with a talent that far exceeds their other abilities (talented savants) or the abilities of the general population (prodigious savants). The syndrome is estimated to affect males about six times as frequently as females. The special skill can appear quite suddenly and disappear equally quickly. Skills commonly displayed by savants include calendar calculations (some can tell the day of a person's birthday in any year of a 1000-year period); mathematical ability; musical ability, including the ability to play new pieces of music after hearing them once; sculpting; drawing; and peculiar feats of memory, such as memory of what the weather was like on every day of the savant's life, retention of the names of all visitors ever received by the savant and the dates of their visits, and the date of every burial in a parish in a 35-year time span as well as the names of all the attendees.

The causes of precocious abilities displayed by cognitively impaired children are not known. Some suggestions are that these children have islands capable of normal function in an otherwise impaired brain, that through some developmental abnormality they are overdeveloped in some brain areas and underdeveloped in others, or that they are using otherwise adequate brains in a functionally unusual manner. Because reasoning is generally poor, the left hemisphere is suggested to be impaired and the right hemisphere intact.

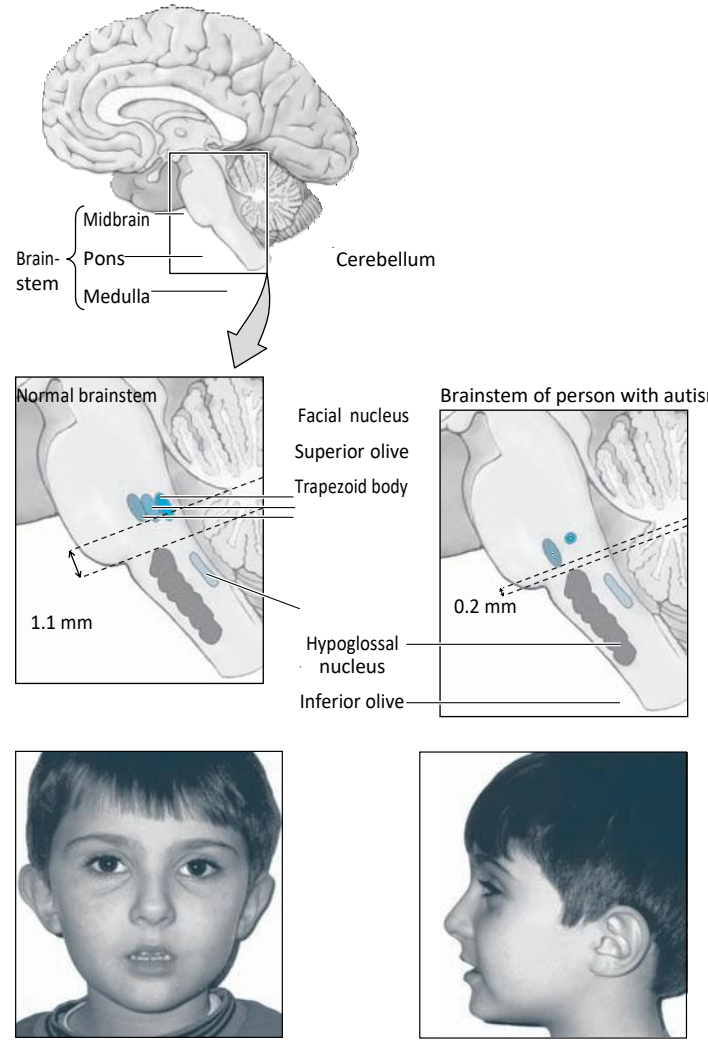


Figure 24.5 Effects of autism. Autism's effects include changes to the brainstem in which the posterior part of the pons is reduced in size. Several nuclei in this region, including the facial nucleus, superior olive, and trapezoid body, are either smaller than normal or missing. A child with autism is normal in appearance but may have some physical anomalies characteristic of the disorder. The corners of the mouth may be unusually low in relation to the upper lip, the tops of the ears may flop over (left), and the ears may be a bit lower than normal and have an almost square shape (right). (Drawings after Roder, © 2000 by Scientific American, Inc., all rights reserved; photographs courtesy of Susan L. Hyman.)

Fragile-X Syndrome

Fragile-X syndrome is the most common inherited cause of mental impairment. It affects about 1 in 2000 males and 1 in 4000 females. About 1 in 259 women and 1 in 800 men carry the fragile-X gene and could pass it on to their children. Fragile-X syndrome is characterized by mental handicaps and facial abnormalities. The mental impairment ranges from subtle learning disabilities to severe mental retardation and is associated with attention deficits, hyperactivity, anxiety and unstable mood, and autistic-like behaviors. The physical abnormalities include a long face, large ears, flat feet, and hyperextensible joints, especially fingers. Boys are typically more severely affected than girls in that most boys with fragile-X syndrome are typically retarded, whereas only about one-third of girls are retarded.

Fragile-X syndrome is caused by an abnormality of the *FMR1* gene, which is located on the long arm of the X chromosome. When functional, this gene encodes a protein (called fragile-X protein) that plays a role in the translation of mRNA into protein in neurons and that plays a role in synapse formation and elimination. The mutation occurs in a stretch of CGG repeats in the DNA. It is a sequence that is prone to increase in length as it is passed from generation to generation. When the number of repeats exceeds a critical level of about 100, the gene no longer makes a functional protein. Examination of neurons in affected persons at autopsy shows that dendritic spines are poorly formed and more numerous. Thus, the protein encoded by the *FMR1* gene may be required for the normal development and elimination of synapses. MRI scans of children with fragile-X syndrome show thinning of the cortex, an unusually small caudate, and an increase in ventricular size. The increase in ventricular size suggests a general loss of brain cells.

The symptoms of females are generally less severe than those of males because females have two X chromosomes; if one chromosome is abnormal, the other is usually able to manufacture the necessary protein. This difference suggests that—in theory at least—if a normal copy of the *FMR1* gene could be inserted into brain cells, neuronal abnormalities could be reduced. Gene insertion has been achieved in neurons cultured in a dish and in a mouse that had previously lacked the fragile-X gene. Another investigational approach to developing a treatment is based on the fact that the gene is capable of producing the protein but is turned off by the excessive number of CGG repeats. Researchers are exploring the possibility of restoring the gene's function without introducing any new genetic material.

Fetal Alcohol Syndrome

The term **fetal alcohol syndrome** (FAS) was coined by Jones and Smith in 1973 to describe a pattern of physical malformation and mental retardation observed in children born of alcoholic mothers. Children with FAS may have abnormal facial features, such as unusually wide spacing between the eyes (Figure 24.6). They also have a range of brain abnormalities, from small brains with abnormal gyri to brains of normal size with abnormal clusters of cells and misaligned cells in the cortex. Related to these anatomical abnormalities are certain behavioral symptoms common to FAS children. They have varying

degrees of learning disability and lowered intelligence scores, as well as hyperactivity and other social problems. They have other physical symptoms, too, including small size and a tendency to be thin.

The recognition of FAS stimulated widespread interest in the effects of alcohol consumption by pregnant women. Pronounced FAS is found in the offspring of approximately 6% of alcoholic mothers. The incidence of it in different geographic regions varies widely, depending largely on the pattern and degree of alcohol abuse in those locations. Streissguth and Connor suggest that about 1 in 700 to 1 in 100 newborns have FAS.

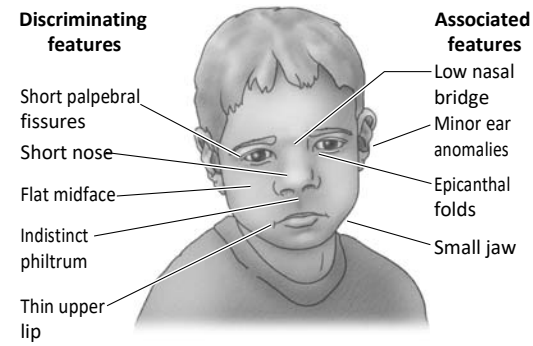


Figure 24.6 Diagram of the characteristics of fetal alcohol syndrome in the face of a young child. Discriminating features, which indicate that the child has fetal alcohol syndrome, can be accompanied by associated features. (After Streissguth and Connor, 2001.)

Fetal alcohol syndrome is not an "all or none" diagnosis. Alcohol-induced abnormalities can range from hardly noticeable physical and psychological effects to the full-blown FAS syndrome. The severity is thought to be related to when, how much, and how frequently alcohol is consumed in a pregnancy. Apparently, the effects are the worst if drinking takes place in the first 3 months, which unfortunately may be a time when many women do not realize that they are pregnant. Severe FAS is also more likely to be caused by binge drinking, which produces high blood-alcohol levels. Other factors related to a more severe outcome are poor nutritional health of the mother and the mother's use of other drugs, including cigarettes.

A major question raised by FAS is how much alcohol is too much to drink during pregnancy. The matter is complex because the effects of alcohol on a fetus depend on so many factors. To be completely safe, it is best not to drink at all in the months preceding pregnancy and during it. This conclusion is supported by findings that as little as one drink of alcohol per day during pregnancy can lead to a decrease in intelligence test scores of children.

Fetal alcohol syndrome in both its full-blown and milder forms has important lessons to teach us. Alcohol is a widely used drug. It poses risks when used inappropriately but, when taken in moderation, is thought to have some health benefits. Even so, it should be avoided completely by women who are pregnant. A major problem is that women who are most at risk for bearing FAS babies are poor and not well educated, with alcohol-consumption problems that predate pregnancy and little access to prenatal care. Often they are unaware of the dangers that alcohol poses to a fetus and do not understand the need to abstain from drinking while they are pregnant.

Developmental Influences on Learning Disabilities

Six factors are most often cited as possible influences on the incidence of learning disabilities: (1) structural damage and toxic effects, (2) hormonal effects, (3) abnormal cerebral lateralization, (4) maturational lag, (5) environmental deprivation, and (6) genetic influences.

Structural Damage and Toxic Effects

When a childhood learning disability resembles a symptom seen in brain-damaged adults (dyslexia is an example), it is only natural to wonder whether the learning disability was caused by structural damage of a similar nature, perhaps resulting from birth trauma, encephalitis, anoxia, or an early-childhood accident. This is no doubt the case for a small minority of children, but many of the neurological symptoms associated with brain damage in adults are not typically observed in children, suggesting that structural damage is not likely to be the cause of most childhood learning disorders. For example, children with developmental dyslexia do not have hemianopsia (blindness in half of the visual field) or scotomas (blind spots in the visual field), symptoms present in a large percentage of brain-damaged adults with dyslexia. Furthermore, the results of EEG and CT-scan studies do not support a structural-damage hypothesis: abnormal EEGs similar to those correlated with known brain damage are not consistently correlated with learning disabilities.

Other possible causal factors include poor nutrition, drug use, and exposure to environmental contaminants. For example, Pihl and Parkes, analyzing specimens of hair from normal children and from learning-disabled children, found significant differences between the groups in the levels of sodium, cadmium, cobalt, lead, manganese, chromium, and lithium. When levels of these same elements were measured in hair taken from a second sample of children, the researchers were able to identify 98% of those who had been diagnosed as being learning-disabled. Although Pihl and Parkes could not identify the cause of the abnormal levels of trace elements, their success in diagnosing learning-disabled children leads to serious consideration of biochemical factors as causal factors in learning disabilities.

The Geschwind-Galaburda Theory

The Geschwind-Galaburda hypothesis proposes that hormones may affect brain development and learning. The seed of this hypothesis lies in Geschwind's observation that the planum temporale (an area thought to represent speech in the left hemisphere) is asymmetrical, being larger on the left and smaller on the right in most right-handers. This asymmetry is thought to be the basis of the underlying neural asymmetry that gives rise to the left hemisphere's dominant role in language. Because males are thought to show greater deviance from this asymmetrical pattern, the possibility that testosterone plays a role is suggested. During embryonic development, the male fetal gonads produce high levels of testosterone, comparable to the levels in adult males. The Geschwind-Galaburda hypothesis proposes that the embryonic surges of testosterone delay the development of the left hemisphere, allowing the right hemisphere both space and time for greater development. Thus, males in general have some comparatively better developed areas in the right hemisphere, which would presumably endow them with excellent spatial skills. If the testosterone-induced asymmetry produces some particularly large right-hemisphere areas, perhaps special abilities, such as precocious mathematical reasoning ability, may result. On the other hand, perhaps testosterone also produces some casualties, characterized by brain abnormalities and learning disabilities. An additional aspect of the hypothesis is that it explains the high incidence of autoimmune disorders (migraines, allergies, asthma, thyroid

disorders, ulcerative colitis, and so forth) both among males in general and among males with exceptional abilities. The hypothesis proposes that testosterone also affects the development of the immune system, with a consequent increase in susceptibility to autoimmune disorders.

The appeal of the Geschwind-Galaburda hypothesis is that it can account for the general observation that females tend to do better than males at language-related tasks and males tend to do better than females at spatial tasks. It also accounts for the high incidence both of precocity and of learning disabilities among males. Furthermore, the proposed shift in cerebral dominance suggests an explanation for the high incidence of left-handedness among the precocious and among the learning-disabled. And, because the effects of testosterone on the brain will in some ways parallel its effects on the immune system, the hypothesis accounts for the high incidence of autoimmune disease in the precocious and learning-disabled male populations. Additionally, the theory allows for deviations in hormonal functions to produce increased incidences of learning disabilities, precociousness, left-handedness, and autoimmune disorders in females. Another appealing aspect is that the hypothesis is testable and can be explored by using animal models.

The first dissection of the brain of a person with a reading disability was performed by Drake, who examined the brain of a 12-year-old boy who died of cerebral hemorrhage. The boy's intelligence had been normal, but in school he had been impaired in arithmetic, writing, and reading. The autopsy revealed atypical gyral patterns in the parietal lobes, an atrophied corpus callosum, and neurons in the underlying white matter that should have migrated to the cortex.

Later Galaburda's group examined the brain of a 20-year-old man who had had a reading disability despite average intelligence. Visual inspection showed nothing abnormal, but microscopic examination revealed several abnormalities. Polymicrogyria (numerous small convolutions) and other architectonic abnormalities were found in the left frontal and parietal cortex. The locations of abnormal brain regions are shown in Figure 24.7. Subcortical abnormalities in the medial geniculate nucleus and the lateral posterior nucleus of the thalamus were discovered, as well. Since the original study, this group has reported similar findings in other cases.

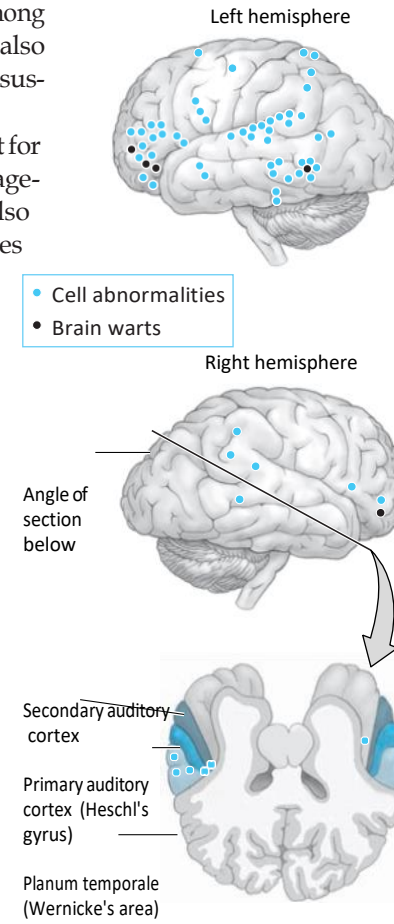


Figure 24.7 Drawings of the two hemispheres, showing the locations of cell abnormalities in the brain of a person who was diagnosed as being reading-disabled. The horizontal section illustrates the asymmetrical pattern of the planum temporale in which the dots, showing areas of cortical anomalies, indicate that the involvement is asymmetrical. (After Geschwind and Galaburda, 1985.)

Abnormal Cerebral Lateralization

A variety of theories rest on Orton's premise that learning disabilities result from slowed cerebral lateralization. This premise is based on the assumption that, because language is lateralized in the left hemisphere of most adults, such lateralization must be advantageous and acquired with language acquisition, and its slowed development would be deleterious to the acquisition of language skills. In the past 20 years, dozens of studies have examined dichotic and visual-field asymmetries, but the data are far from unequivocal. Satz concludes:

> One might ask what light laterality studies shed, if any, on the problem of cerebral dominance and reading disability. The answer should be — not much. The reason for this somewhat discouraging view lies in the numerous methodological and conceptual problems that continue to plague research efforts in this area. (Satz, 1976, p. 288)

Maturational Lag

The maturational-lag hypothesis postulates that the cognitive functions producing language, reading, and other complex behaviors are organized hierarchically and that the levels of the hierarchy develop sequentially in the course of ontogeny. Should one level of the hierarchy be slow to develop, the development of all subsequent levels will be delayed, inasmuch as higher functions depend on the integrity of lower ones. The original delay in maturation could result from a variety of factors. Two possibilities are delayed myelinization of a particular region and slow development of cortical connections.

Although the results of some studies do suggest that various functions in learning-disabled children are slow in maturing, the type of study needed for a definitive test of this hypothesis is a careful longitudinal analysis of children tested on a large number of perceptual, motor, and cognitive skills for a period of 10 to 15 years. When learning-disabled children *have* been reexamined in adulthood, they have been found to retain their characteristic impairments even though maturation should have been complete. For example, Frauenheim studied 40 adults who had been diagnosed as dyslexic in childhood and found that, on test performance and on self-report evaluations, they were essentially unchanged from the original diagnosis. This result does not support the maturational-lag hypothesis.

Environmental Deprivation

It is well known that environmental deprivation can have long-lasting consequences for physical and intellectual function. Children raised in orphanages with adequate physical care but without adequate social stimulation fail to thrive. They fall below age norms in both physical and intellectual development. A recent example is that of Romanian orphans. In the 1970s, the communist regime then governing Romania outlawed all forms of birth control and abortion. The result was hundreds of thousands of unwanted pregnancies, with children placed in orphanages, where the conditions were appalling. After the Communist government fell and the outside world was able to intervene, hundreds of these children were placed in adoptive homes throughout the world. There have been several studies of the fate of these children. Initially malnourished and small in size, they improved spectacularly in their adoptive homes. Their average height and weight became almost normal, and most achieved normal motor and cognitive development. A significant number were retarded, however, and many had psychosocial problems—difficulty in developing relations with peers and in developing secure attachments with adults. Children who were adopted before 6 months of age had significantly better outcomes than did those adopted at older ages.

People in developed countries continue to adopt children from developing countries—children who have been subjected to various degrees of deprivation. In addition, many children in even the most advanced countries suffer various degrees of deprivation and abuse. Accordingly, environmental deprivation continues to be a leading cause of learning disabilities.

The Birthday Effect

A subtle variant of the deprivation hypothesis is called the **birthday effect.** One perspective on this hypothesis comes from studies undertaken by Barnsley and colleagues of birthdays of North American hockey players. In senior hockey leagues, there is a negative relation between birth month and number of players. More than 30% of players have birth dates in the first quarter of the year (16% in January), whereas fewer than 15% have birth dates in the last quarter of the year (5% in December). Furthermore, a disproportionate number of the superstars have first-quarter birthdays. This birth discrepancy is not present in beginning leagues but emerges progressively as players are promoted through the leagues. The explanation appears to be straightforward. Players enter the most junior league according to age—children must be 8 years old between January 1 and December 31 of the year in which they enter Mite hockey. Equal numbers of children born in each month enter. But children born in December enter hockey almost a year earlier than children born in January, who in effect have had to wait a year. The younger, smaller children are at a developmental disadvantage from the outset. They receive less playing time and reinforcement and are more likely to drop out.

Research on the effects of relative age on educational achievement produces similar results. Children entering school at a younger age perform at a significantly lower level than do their older classmates. Both Diamond and Maddux found that children entering first grade at an early age were more likely than their older classmates to be classified as learning-disabled later in their school career. Furthermore, Maddux and coworkers found that, among children who are classified as gifted, a larger population entered school late than entered school early. This effect may last into later grades and even into university. The rather simple birthday effect stands in sharp contrast with the brain-based hypotheses and should inspire sober reflection in neuropsychologists who are making diagnoses.

Genetic Bases of Learning Disabilities

Any consideration of the possibility that learning disorders have a genetic basis must recognize some of the obstacles to demonstrating such a hypothesis. First, the environment can affect development in many ways. Consequently, it is extremely difficult to separate environmental influences from genetic effects in any kind of research. Second, learning disabilities take many forms. At present, criteria for categorizing types of learning disabilities are poorly developed, which makes it difficult for researchers to correlate specific types of disabilities with specific causes. Third, the incidence of learning disabilities is related to the quality of schooling. The average length of schooling and the demands made by schools on students have changed greatly in the past two generations; so it is difficult to compare the reading abilities of children with those of their parents. Fourth, learning-disabled children are typically of average intelligence, as are their parents (that is, their overall IQs are about 100), and people with strictly average IQs generally find school difficult, even when no specific disability exists. Fifth, the ability to read is itself probably inherited, making it difficult to sort out the contribution made by inherited reading skill from that made by a supposedly inherited causal factor underlying a disability.

Despite the difficulties of obtaining meaningful research results, the possibility of genetic causes has been raised repeatedly. Since the early twentieth century, many authors have referred to the high incidence of learning disabilities within certain families, and studies of twins have found a higher incidence of dyslexia in identical compared with fraternal twins. Asherson and Curran note that many genes are likely implicated in learning disorders. The results of different studies of families with a high incidence of reading disabilities suggest that there may be genes on chromosomes 1, 2, 6, and 15 related to the disorder. As our understanding of genes and their influence advances, it is quite possible that hundreds rather than just a few genetic abnormalities will prove to be associated with learning disabilities.

The Adult Outcome of Learning Disabilities

There are a variety of views about the outcome of children with learning disabilities. In the most optimistic outcome study, reported by Critchley, 20 dyslexic boys attended a private school and received special instruction using training methods. As adults, two of the boys became medical doctors, two college professors, one a lawyer, two research scientists, six owners or managers of businesses, one a school principal, three teachers, one an actor, one a factory foreman, and one a skilled laborer. There are also reports in the popular press of similar, though perhaps not so absolute, successes at various private schools for learning-disabled children. Most studies do not report such optimistic outcomes, however, and the most thorough of them reaches frankly pessimistic conclusions concerning academic outcome. Spreen examined the progress of 203 learning-disabled people over a long period, deriving the following findings from assessments, personal interviews, parental interviews, and other observations and data:

1. Persons in all but the control group suffered through a miserable and usually short school career and then experienced a miserable social life full of disappointments and failures. They also had a relatively poor chance of obtaining advanced training and skilled employment. They did not, however, have a higher incidence of juvenile delinquency or psychiatric problems.
2. Interviewed separately, subjects and their parents were largely in agreement concerning factual information, but parents tended to regard the learning disabilities as having had more serious effects on the well-being, happiness, and social interaction of their children than were reported by the children themselves. The affected children also had less-detailed memories of their childhood than the control children did. As the subjects aged, they developed firmer plans for their future and made better occupational adjustments, but they also gave increasingly negative descriptions of their school experiences. The eventual social adjustments of the females were worse than those of the males.
3. Although the learning-disabled eventually managed to make personal adjustments and find jobs, their dislike of school and dissatisfaction with it persisted.

Weiss and Hechtman reported that the prognosis for hyperactive children is relatively poor. Their behavior continues to be impulsive, and their lives are often characterized by frequent relocations, accidents, problems with the law, and so forth. As adults, their major problems may be low self-esteem and poor social skills, but they are not regarded as hyperactive by their employers, and they do not become mentally disturbed or commit crimes.

We end this section by emphasizing the importance of careful assessment in evaluating the particular cognitive deficits of each learning-disabled child. After problem areas have been identified, specialized teaching programs can be devised to circumvent handicaps. There may be little point in trying to teach a given child a particular skill that he or she is clearly not ca- pable of learning. Perhaps the educational program for that child should in- stead be directed toward the acquisition of skills that can be used to gain employment. Counseling is an important part of the educational process both for the learning-disabled child and for the parents. It should focus not only on overcoming negative attitudes toward the educational system but also on understanding the child's unique handicaps and on devising strate- gies for circumventing some of them.

Summary

A variety of disorders appear in childhood and interfere with progress in school and in social adjustments. The acquisition of reading is central in school and so disorders that result in impairments in reading are understandably an obstacle to satisfactory academic progress. Although reading is a complex ac- tivity that can be disrupted in many different ways, a growing body of evidence suggests that an ability to make rapid discriminations and attentional shifts is particularly important for acquiring literacy. Procedures that specifically train subjects in stimulus discrimination have therefore proved helpful.

A number of common nonverbal disabilities lead to academic and social dif- ficulties. Many of these conditions are associated with general diffuse damage that varies from case to case. A variety of environmental conditions also can in- fluence brain function and developmental success, including brain injury, tox- ins, drugs, and environmental deprivation. Even somewhat more subtle influences such as the age at which a child begins school can have a surprisingly large effect on school success.