Disorders of Visual Perception

Because the visual system is somewhat modular, damage to a processing area can impair one aspect of visual perception while all others remain normal. This kind of deficit is often called an *agnosia*, which means "lack of knowledge." Because the disorders provide a special opportunity for understanding the neural basis of higher-order visual perception, we will orient our discussion of the perception of objects, color, movement, and spatial location around disorders of those abilities.

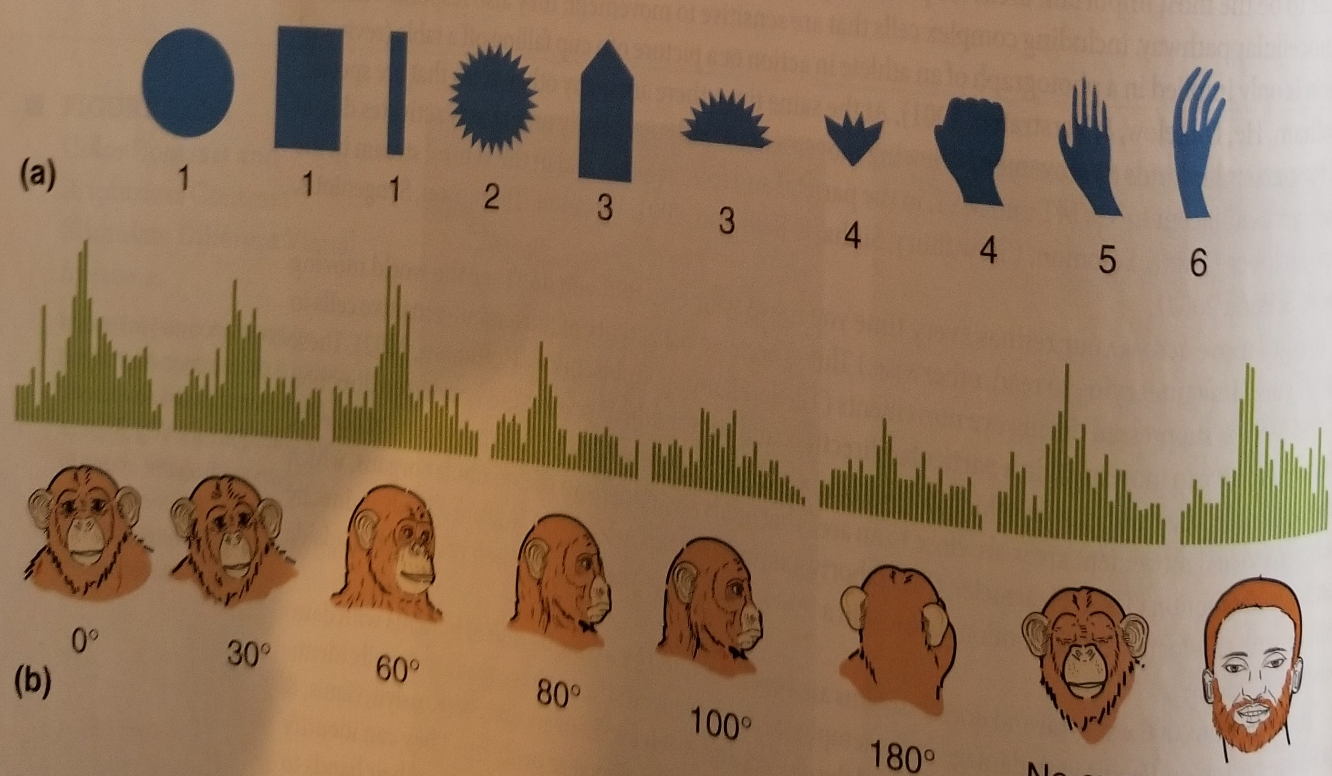
Object and Face Agnosia

Object agnosia is the impaired ability to recognize objects. In Chapter 3, we described Oliver Sacks's (1990) agnosic patient who patted parking meters on the head, thinking they were children; he was also surprised when carved knobs on furniture failed to return his friendly greeting. Dr. P. was intellectually intact; he continued to perform successfully as a professor of music, and he could carry on lively conversations on many topics. Patients with object agnosia are able to see an object, describe it in detail, and identify it by touch. But they are unable to identify an object by sight or even to recognize an object from a picture that they have just drawn from memory (Gurd & Marshall, 1992; Zeki, 1992).

Object agnosia is caused by damage to the inferior temporal cortex (see Figure 10.29); this part of the ventral stream is where information about edges, spatial frequencies, texture, and so on is reassembled to form perceptions of objects. Cells have been located there in monkeys and humans that respond selectively to geometric figures, houses, animals, hands, faces, or body parts (Figure 10.30a; Desimone,

FIGURE 10.30 Stimuli Used to Produce Responses in Hand- and Face-Sensitive Cells in Monkeys.

(a) The stimuli are ranked in order of increasing ability to evoke a response in a hand-sensitive cell. (b) The spikes recorded from a face-sensitive cell indicate the degree of response from the stimulus shown below.



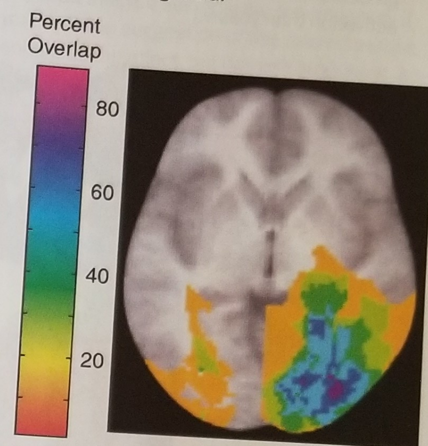
Sources: (a) From "Visual Properties of Neurons in Inferotemporal Cortex of the M... with permission. (b) From "Stimulus-Selective Properties of Inf... 4, 1984.

Albright, Gross, & Bruce, 1984; Downing, Jiang, Shuman, & Kanwisher, 2001; Gross, Rocha-Miranda, & Bender, 1972; Kreiman, Koch, & Fried, 2000; Sáry, Vogels, & Orban, 1993). Some of these cells require very specific characteristics of a stimulus, such as a face viewed in profile; others continue to respond in spite of changes in rotation, size, and color (Figure 10.30b; Miyashita, 1993; Tanaka, 1996; Vogels, 1999). The latter group of cells likely receive their input from cells with narrower sensitivities (Tanaka, 1996), like those in V1 that detect edges. The inferior temporal cortex also has a columnar organization reminiscent of what we saw in V1; a column of object-responsive cells might respond to variations on a star-like shape, for example, and a column adjacent to one that responds to a frontal view of a face is activated by a face in profile (Tanaka, 2003).

Like Dr. P., many object agnosic patients also suffer from *prosopagnosia*, an impaired ability to visually recognize familiar faces. The problem is not memory, because they can identify individuals by other characteristics like their speech or mannerisms. Nor is their visual acuity impaired; they often have no difficulty recognizing facial expressions, gender, and age (Tranel, Damasio, & Damasio, 1988). However, they are unable to recognize the faces of friends and family members or even their own image in a mirror (Benton, 1980; A. R. Damasio, 1985). Prosopagnosia has a variety of causes, including stroke, carbon monoxide poisoning, and Alzheimer's disease. Damage usually impairs the ability to recognize both objects and faces, but the occasional case is reported of a patient with prosopagnosia alone (Benton, 1980) or of object agnosia with spared face identification (Behrmann, Moscovitch, & Winocur, 1994).

FIGURE 10.31 Location of Brain Damage in Patients With Prosopagnosia.

The color of the area indicates how often damage was observed there in patients with prosopagnosia.



Source: From "Behavioral Deficits and Cortical Damage Loci in Cerebral Achromatopsia," by S. E. Bouvier and S. A. Engel, 2006, *Cerebral Cortex, 16*, pp. 183–191, by permission of Oxford University Press.

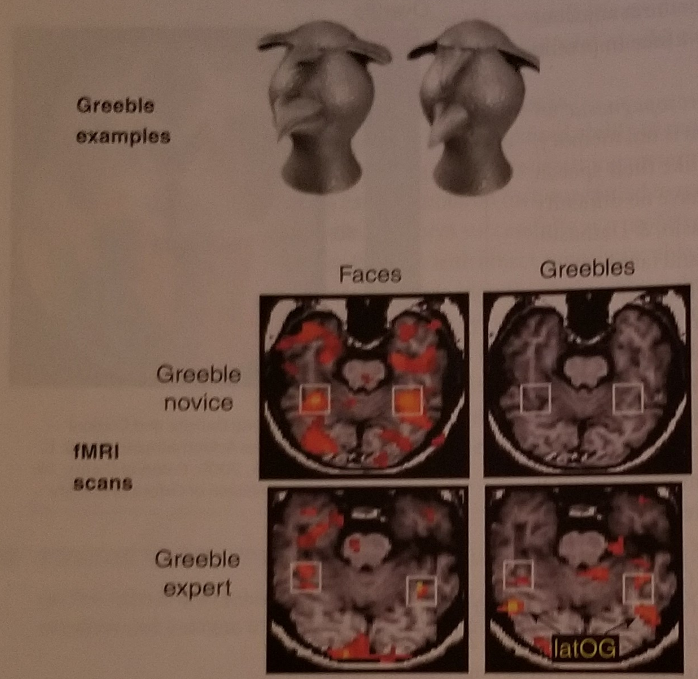
While some processing of face information occurs in the inferior temporal cortex, recognizing individual faces requires additional structures. In humans, a part of the fusiform gyrus on the underside of the temporal lobe is so important to face recognition that it is referred to as the *fusiform face area (FFA)*. This area starts assisting us in recognizing caregiver faces at about two years of age (Bushnell, 2001) but can be developmentally delayed in individuals with autism (Scherf, Behrmann, Minshew, & Luna, 2008). Damage that results in prosopagnosia is usually in the right hemisphere (Figure 10.31; Bouvier & Engel, 2006; Gauthier, Skudlarski, Gore, & Anderson, 2000; Gauthier, Tarr, Anderson, Skudlarski, & Gore, 1999), but face processing is a cooperative effort involving both sides of the brain. Facelike images produce activity in the left fusiform gyrus that strengthens as the resemblance to a face increases; about two seconds later, the right fusiform gyrus increases its activity only when the image is of a human face (Meng, Cherian, Singal, & Sinha, 2012). Another area that has been implicated in face recognition is the medial temporal lobe, which is important in memory and seems to be involved in decision making between familiar and unfamiliar faces, even when features are shared between those two groups of faces (Quian Quiroga, Kraskov, Mormann, Fried, & Koch, 2014).

Until recently, researchers thought the only way prosopagnosia occurred was through brain damage. Then medical student Martina Grueter began to recognize the symptoms in her husband's behavior and made congenital prosopagnosia the subject of her MD thesis (Grueter, 2007). An estimated 2.5% of the population has symptoms of the disorder without any history of brain damage (Kennerknecht et al., 2006); thus, they make errors in recognizing familiar faces, and they learn new faces slowly (Grüter, Grüter, & Carbon, 2008). Afflicted individuals include noted primatologist Jane Goodall, actor Brad Pitt, and Oliver Sacks, the neurologist who studied Dr. P. Face recognition ability has a heritability of about 39% (Zhu et al., 2010), so its deficiency in the absence of brain damage has a genetic origin. The defect, though, does not appear to be in the fusiform face area; fMRI shows that the FFA responds just as much to faces in prosopagnosics as it does in normal individuals. Instead, connections of the FFA to more anterior temporal and frontal cortex areas are diminished (Avidan & Behrmann, 2009), suggesting that face recognition is a distributed function, despite modularity of its components.

These capabilities might be "hardwired" at birth to some extent, but they also are amenable to learning. When researchers showed monkeys pictures of the faces of lab workers, neurons in the inferior temporal cortex increased their firing rates according to the monkeys' familiarity with the workers (M. P. Young & Yamane, 1992). Isabel Gauthier and her colleagues (1999) trained humans to identify faces, using pictures

FIGURE 10.32 Activity in the Fusiform Face Area While Viewing Faces and "Greebles."

Viewing faces activated a part of the fusiform gyrus (indicated by the white squares) both in "greeble novices" and in "greeble experts," who had learned to distinguish individual greebles from each other. Viewing greebles activated the area only in greeble experts. (Red indicates more activation than yellow.)



Source: From "Activation of the Middle Fusiform 'Face Area' Increases With Expertise in Recognizing Novel Objects," by I. Gauthier et al., *Nature Neuroscience*, 2, pp. 568–573. © 1999 Nature Publishing Group.

of fictitious creatures they called "greebles" to ensure initial unfamiliarity. The fMRI scans in Figure 10.32 show that pictures of human faces activated the FFA but greebles did so only after the person had learned to recognize individual creatures.

Prosopagnosics do respond emotionally to photographs of familiar faces they do not recognize, as indicated by EEG evoked potentials and skin conductance response (Bauer, 1984; Renault, Signoret, Debruille, Breton, & Bolger, 1989; Tranel & Damasio, 1985). This "hidden perception" is not without precedent. Patients blinded by damage to V1 show a surprising ability to track the movement of objects and discriminate colors, all the while claiming to be guessing (Zeki, 1992). Cortically blind individuals also can identify emotions expressed in faces they do not otherwise see (Tamietto et al., 2009), and they can avoid obstacles while walking. This ability to respond to visual stimuli that are not consciously seen is called *blindsight*. Imaging studies have found that blindsight depends on pathways passing through the superior colliculus directly to extrastriate areas, bypassing V1 (reviewed in de Gelder & Tamietto, 2007; Tamietto et al., 2010).

A nearby area in the inferior temporal cortex has an intriguingly similar "object" recognition function; the *visual word form area (VWFA)* responds to written words as a whole. Its importance in reading was demonstrated in a patient whose VWFA was disconnected from adjacent language areas by surgery intended to remove tissue that was causing epileptic seizures (Gaillard et al., 2006). Before surgery, the patient could recognize familiar words of any length in less than a second; following surgery, the time had almost doubled for three-letter words and increased by an additional 100 milliseconds for each additional letter, indicating that he was deciphering words letter by letter. Performance in identifying faces, tools, and houses was unaffected. The VWFA is typically underactivated in adult dyslexics during reading (McCandliss & Noble, 2003), but, consistent with what we saw in Chapter 9, the authors suggest that this is not the cause of the dyslexia but the result of phonological deficits that interfere with learning rapid word recognition. It is clear that the VWFA could not have evolved as a dedicated whole-word detector, because written language is a relatively recent invention; still, it serves that function so precisely that two words evoke activity in different subareas, even when the words differ by just one letter (Glezer, Jiang, & Riesenhuber, 2009). What is intriguing about the VWFA is that, for whatever reason, the area has evolved special capabilities that suit it for learning to identify words as if they are unique visual "objects."

> "I was having a wonderful conversation with a woman at a party, but then I went to get us some drinks. When I returned, I had forgotten what she looked like, and I was unable to find her the rest of the evening."
>
> —A young man with prosopagnosia

Color Agnosia

? How is color coding different from wavelength coding?

Let's return to Jonathan I., whose plight was described at the beginning of the chapter. Jonathan's problem was *color agnosia*, which is the loss of the ability to perceive colors due to brain damage. But before we can discuss this disorder, we need to revisit the distinction between wavelength and color. As one of the authors walked past a colleague's slightly open office door, he was astonished to see that his colleague's face was a distinct green! Opening the door to investigate, he understood why: The light from the colleague's desk lamp was reflecting off a bright green brochure he was reading. Immediately his face appeared normal again. This ability to recognize the so-called natural color of an object despite the illuminating wavelength is called *color constancy*. An example of this can be seen in Figure 10.33. Although both eyes are the same color gray, the colored shading pushes the brain's interpretation of color in the opponent direction. If not for color constancy, objects would seem to change colors as the sun shifted its position through the day or as we went indoors into artificial (or colored) light. Imagine having to survive by identifying ripe fruit if the colors kept changing.

When the author reinterpreted his colleague's skin color, it was not because he *understood* that his friend's face was bathed in green light; it occurred automatically as soon as his eyes took in the whole scene. Monkeys, who do not understand the principles of color vision, apparently have the same experience. When Zeki (1983) illuminated red, white, green, and blue patches with red light, each patch set off firing in V1 cells that preferred long-wavelength (red) light, regardless of its actual color; however, cells in V4 responded only when the patch's actual color matched the cell's color "preference." Zeki concluded that cells in V1 are *wavelength coded*, whereas cells in V4 are *color coded*. Schein and Desimone (1990) suggested how V4 cells provide color constancy. They have large circular receptive fields that are color opposed; so if, for example, a green light falling on the center increases the cell's firing rate, green light falling simultaneously in the surround reduces or eliminates the increase. In other words, the cells "subtract out" the color of any general illumination. It is our V4 cells that allow us to see our friend's face as normal pink rather than as the color that it might reflect.

FIGURE 10.33 Color Constancy

In both images, all the eyes are gray. However, our cortex corrects for the right-sided tint; it tells us what color eye would produce the sensation we're experiencing, if it were seen through a blue or red glass. Punch a small circle in a piece of paper so that you see only the eye and verify it yourself! The suggestion of the colored earring enhances the effect, but doesn't produce it, as you can see if you cover the earring.

We have no brain scan to tell us where Jonathan I.'s damage was located, but we know that cortical color blindness, or *cerebral achromatopsia*, occurs when people have lesions between V1 and the fusiform face area (Heywood & Kentridge, 2003; Witthoft et al., 2013); this is where V4 is located, but it is unclear whether the deficiency can be attributed to V4 malfunction. Unlike Jonathan I., many patients are unaware their color vision is impaired, just as we saw in Chapter 9 with Wernicke's aphasia.

Movement Agnosia

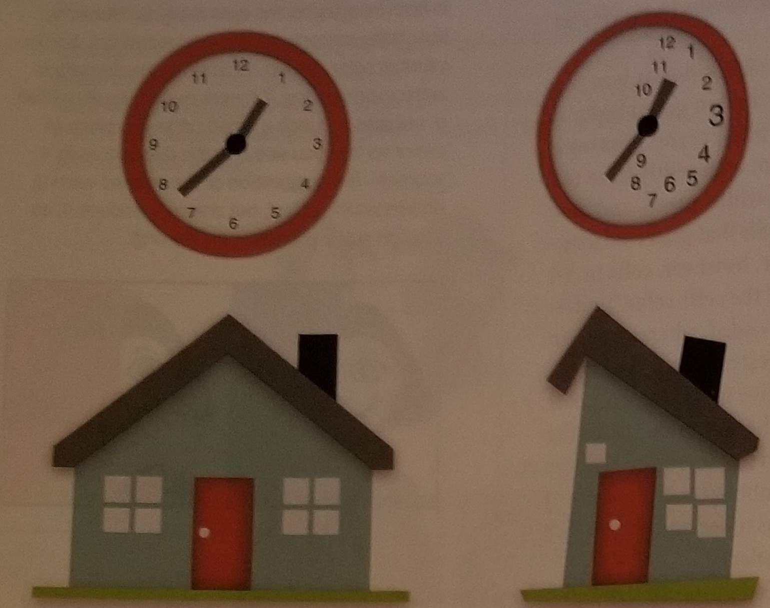
Although movement is detected by neurons in V1 and beyond, area V5/MT is the place where that information is integrated; MT (for middle temporal area) also helps direct reaching movements and eye movements when tracking objects (Born & Badley, 2005; Whitney et al., 2007). A 43-year-old woman known in the literature as LM suffered a stroke that caused bilateral damage in the general area of MT; the result was *movement agnosia*, an impaired ability to detect movement (Vaina, 1998; Zihl, von Cramon, & Mai, 1983). Although her vision was otherwise normal, she could distinguish between moving and stationary objects only in her peripheral view, she had difficulty making visually guided eye and finger movements, and she had trouble detecting the movement of people if there were more than two people in the room. She was often surprised to notice that an object had changed position (Zihl et al., 1983). You might think that perceiving a change in position would be the same thing as perceiving movement, but she had no sense of the object traveling through the intermediate positions. When she poured coffee, she could not tell that the liquid was rising in the cup, so she would keep pouring until the cup overflowed! When she tried to cross a street, a car would seem far away, but then suddenly very near.

Later analyses indicated that LM's most severe impairment was in her ability to detect radial movement (Vaina, 1998). We experience radial movement when the image of an approaching car expands outwardly, or radially. Radial movement also tells us that we are approaching an object when we walk or drive, because all the environmental objects around the central point appear to move outward; this effect provides information about our *heading* and is important for personal navigation. A patient with impaired perception of radial movement could not catch a ball that was thrown to him, and he frequently bumped into people in his wheelchair. Scans done while subjects perform a task involving radial movement or heading detection implicate the MST area (for medial superior temporal area, where it is located), which receives its input from MT (Peuskens, Sunaert, Dupont, Van Hecke, & Orban, 2001; Vaina).

Neglect and the Role of Attention in Vision

The posterior parietal cortex combines input from the visual, auditory, and somatosensory areas to help the individual locate objects in space and to orient the body in the environment. Damage impairs abilities such as reaching for objects, but it also often produces *neglect*, in which the patient

■ **FIGURE 10.34** Drawings Copied by a Left-Field Neglect Patient.



Source: Schwartz, B. L. & Krantz, J. H. (2015).

ignores visual, touch, and auditory stimulation on the side opposite the injury. The term *neglect* seems particularly appropriate in patients who ignore food on the left side of the plate, shave only the right side of the face, or fail to dress the left side of the body. The manifestations are largely, but not entirely, visual, and they are more likely to occur on the left side of the body, following right-hemisphere damage. (Because the symptoms affect one side of space, the term *hemispatial neglect* is often used.)

Neglect is not due to any defect in visual processing, but rather it is due to a deficit in attention; it illustrates the fact that to the extent attention is impaired, so is visual functioning. Two patients with this condition, caused by right parietal tumors, were asked to report whether words and pictures presented simultaneously in the left and right visual fields were the same or different. They said that the task was "silly" because there was no stimulus in the left field to compare, yet they were able to answer with a high level of accuracy (Volpe, LeDoux, & Gazzaniga, 1979). Their performance is superior to that of blindsighted individuals, which supports the contention that neglect is a deficit in attention rather than in vision.

> *"I knew the word "neglect" was a sort of medical term for whatever was wrong but the word bothered me because you only neglect something that is actually there, don't you? If it's not there, how can you neglect it?"*
>
> *—P. P., a neglect patient*

Patients' drawings and paintings help us understand what they are experiencing. When asked to copy drawings, they will neglect one side while completing the other side in detail, like the example in Figure 10.34. The two portraits in Figure 10.35 were painted by Anton Raderscheidt two and nine months after a stroke that damaged his right parietal area. Notice that the first painting has very little detail and the left half of the image is missing. In the later painting, he was using the whole canvas, and the portrait looks more normal; but notice that the left side is still much less developed than the right, with the eyeglasses and face melting into ambiguity (Jung, 1974).

■ **FIGURE 10.35** Self-Portraits Demonstrating Left Visual Field Neglect.

A self-portrait done two months after the artist's stroke, which affected the right parietal area, is incomplete, especially on the left side of the canvas. (b) One done nine months after the stroke is more complete but still shows less attention to detail on the left side.