AGNOSIA

AGNOSIA is a relatively rare neuropsychological symptom defined in the classical literature as a failure of recognition that cannot be attributed to elementary sensory defects, mental deterioration, attentional disturbances, aphasic misnaming, or unfamiliarity with external stimuli (Frederiks, 1969). Agnosia is most often modality-specific; the patient who fails to recognize material presented through a particular sensory channel (e.g., visual, auditory, or tactile) has received the most attention, and will be reviewed here.

One of the most fundamental and difficult questions about agnosia concerns whether it is best thought of as a perceptual or neuronal impairment. In his classic definition, Teuber (1968) stated that “two limiting sets of conditions: failure of processing and failure of naming...” and then proceeded to describe the alleged disorder of recognition per se, which would appear in its purest form as a normal percept that has somehow been stripped of its meaning.” Teuber’s definition is significant since it locates the agnosia at the intersection of perception and memory, and in fact, seems to imply that “pure” agnosia is a disorder of memory access. In contrast, others have asserted that such a deficit does not exist and that all so-called agnosias are either perceptually impaired, or demented, or both (Bay, 1955; Bender and Feldman, 1972). Proponents on either side of this debate assume that perception and memory are dissociable. As we shall discuss, this assumption derives from serial information-processing models of the perceptual process, and alternative models that do not require such a separation are now available (McClelland and Rumelhart, 1986; Rumelhart and McClelland, 1986).

An early observation of agnosia-like phenomena was provided by Munk (1851), who observed that dogs with bilateral occipital lobe excisions nearly avoided obstacles placed in their paths, but failed to react appropriately to objects that previously had frightened or attracted them. Similar observations have been made by Horel and Keating (1960, 1972) in the macaque with lesions of the occipital lobe and its temporal projections. Munk felt that his dogs’ behavior resulted from a loss of memory images of previous visual experience and termed the phenomenon “agnosia.” Nine years later, Lissauer (1890) provided the first detailed report of a recognition disturbance in humans, and his views on different varieties of the disturbance have had important historical impact on theory and practice. The term “agnosia” was introduced by Freud (1891), eventually replacing “mind blindness” and other terms such as “asymbolia” (Flanellburg, 1870) and “imperception” (Jackson, 1879). As with most neurobehavioral syndromes, significant debate has existed regarding the functional mechanisms responsible for agnostic phenomena. However, unlike most other neuropsychological phenomena, a major point of debate has centered on whether agnosia existed at all. Over the years, interpretation of agnostic syndromes has varied according to the zeitgeist prevailing at the time. In the early 20th century, when Gestalt psychology guided perceptual theory, published cases of agnosia were conceptualized with Gestalt concepts in mind (see Goldstein and Gell, 1915; Goldstein, 1943, vs. Poppelreuter, 1923; Brain, 1941). With the reemergence of “disconnection theory” (Geschwind, 1965), cases of agnosia during the 1960s and 1970s were largely viewed as examples of sensory-axon or sensory-mark connections. In the past decade, with significant advances in computational neuroscience and an increased component understanding of visual cognition, clinical data have been reinterpreted in the language of cognitive neuropsychology. The history of agnosia contains several striking examples of the interplay between cognitive theory and clinical practice, and represents a good example of how scientific advancement is not always linear or cumulative. There has been a recent revolution in the field of agnosia as we have moved primarily from an almost exclusive emphasis on disconnection concepts to a more cognitive neuropsychological perspective. Because models of normal perception have always driven conceptualizations of agnosia, a brief review of four broad models will be provided before discussing the major agnostic syndromes.

MODELS OF RECOGNITION

STAGE MODELS

The earliest neuropsychological ideas of the process of object recognition were embodied in “stage models,” which held that the cortex first built up a percept from elementary sensory impressions. Recognition was achieved in a subsequent stage in which the resulting per-
from the syndrome of visual object agnosia, which, in his view, was most often seen in the context of left medial occipital lobe damage. According to Geschwind, this lesion not only induced a right homonymous hemianopia but also prevented information perceived by the intact right hemisphere from reaching the naming area because of impairment of crossing fibers. In advancing this hypothesis, Geschwind (1965) described several examples of patients who, after failing to identify objects on formal testing, later used or interacted normally with the object. In bringing attention to these phenomena, Geschwind (1965) provided clear evidence that recognition is not a unitary phenomenon. He wrote:

A fundamental difficulty has been in the acceptance of a special class of defects of recognition, lying somewhere between deficits of "perception" and "memory". What indeed are the criteria for recognition and is it a single function? I believe that there is no single faculty of recognition, but that the term covers the totality of all the associations aroused by any object. Phrased another way, we "manifest" recognition by responding appropriately, to the extent that any appropriate response occurs, we have "recognized". Not that this view establishes the notion of a unitary step of "recognition"; instead, there are multiple parallel processes of appropriate response to a stimulus. To describe the behavior correctly we must describe the pattern of loss and preservation of responses to each particular type of stimulus. (p. 557)

Although this idea is critical to an understanding of agnosia, it is now clear that disconnection theory itself, accounts for the fact that most agnosics show abnormal verbal and nonverbal processing of viewed objects. Despite this, the major historical impact of this idea was to point out the fact that recognition behavior measures the output of many separate components. Thus, an answer to the basic question, "did the patient recognize?", depends on what response is required of the patient in a given task situation.

COMPUTATIONAL MODELS

The models proposed by Lissauer and Geschwind attempt to explain agnostic symptoms in terms that were consistent with available theoretical constructs. An alternative approach is to begin by accounting for normal perceptual phenomena and to then determine whether such an account can explain recognition failures observed in the clinic. This approach begins by specifying the tasks that sensory-perceptual systems must perform to achieve the kind of powerful and flexible recognition abilities we as humans possess. We are able to recognize everyday objects and faces with remarkable ease across wide ranges in viewing distance, orientation, and illumination. We are able to "infer" depth, volume, and structure from relatively impoverished two-dimensional stimuli such as photographs and line drawings. We can determine with intimate certainty whether a pictured and real object are the same or different, or whether they would be used together. Thus, from perceptual analysis, we can derive an enormous amount of structural and semantic information about the world around us. What is required to perform all these remarkable functions is an ability to answer this question, Marr (1982) started with the assumption that the brain must store some form of coded, symbolic description (a "representation") of known objects or faces that is sufficiently flexible to accommodate the perceptual variations inherent in everyday recognition tasks. His analysis led him to distinguish three types of representations, which he referred to as (1) the primal sketch, (2) the viewpoint-centered, or 2½-D, sketch, and (3) the object-centered, or 3-D, sketch. The primal sketch represents intensity (brightness) changes across the field of vision, resulting in a way of specifying the two-dimensional-approximation of the shape of the image. The 2½-D sketch represents the spatial locations of visible surfaces from the point of view of the observer. The essential feature of this type of representation is that it is computed on the basis of the spatial relationship between viewer and object. Because of this, the resulting representation is dependent on the point of view, and thus it is known as a "viewer-centered" or "viewpoint-dependent" description. The 3-D sketch specifies the configuration of surfaces, features, and shapes within an object in an object-centered coordinate frame (in which shapes and features are represented in terms of their location on the object) yields a description that is not dependent upon the observer's point of view since, for example, simple rotation would not alter the spatial relationships among features of the object. Because of this, the 3-D representation has also been referred to as an "object-centered" or "viewpoint-independent" description. Presumably, achieving this kind of description is essential to flexible object recognition, although it is obvious that specific objects could sometimes be recognized using only a 2½-D sketch (see below).

Marr's theoretical position is important for two reasons. First, it provides an a priori conceptual approach to the study of object recognition disturbances. Indeed, we will see that Marr's ideas about multiple object representations provide a useful framework within which to understand the various ways that recognition can become disordered. Marr's ideas have led to the development of new, more refined, clinical assessment tools, and have clarified some of the intractable problems in this area. Second, it serves as a potent reminder that the various tests used to tap the apperceptive and associative stages of object recognition impose different demands on the recognition apparatus.

In a contemporary computational model set in neural terminology has been proposed by Damasio (1989). Like its ancestors in the parallel-distributed processing framework (McClelland and Rumelhart, 1986; Rumelhart and McClelland, 1986; Goldman-Rakic, 1985), Damasio's model suggests that perception involves the retrieval activity pattern in primary and first-order association cortex that corresponds to the various perceptual features extracted from viewed objects. Downstream, these features are combined in "local convergence zones," which serve to bind together the pattern of features into an "entity" (e.g., object). Damasio specifically rejects the view that recognition involves the activation of a packaged, locally stored memory representation of the stimulus. Instead, recognition occurs when the neural pattern defining a specific entity is reactivated in a time-locked fashion in retrospective. The most important feature of Damasio's model is that there is no fundamental distinction between perception and memory is made; that is, information about previously encountered items is stored in a pattern of neural activity, not in a localized representation. In this sense, recognition is "re-cognition." Because the memory-perception distinction is abolished, this kind of model avoids many of the problems encountered in answering the question, "is agnosia a perceptual or memory deficit?" Damasio's model predicts that there can be no disconnection of object recognition without attendant perceptual dys-function. As we shall see, this seems entirely consistent with the behavior of most associates.

COGNITIVE NEUROPSYCHOLOGICAL MODELS

A fourth class of models has recently emerged in the tradition of the cognitive modular, or "box model," approach. These models attempt to outline, in cognitive terms, the functional components involved in object recognition. Such models have received significant attention only in the context of visual recognition. One representative model of object recognition, proposed by Ellis and Young (1988), is depicted in Figure 13–1. In this model, the initial, viewer-centered, and object-centered representations correspond to Marr's three levels of object description. According to Ellis and Young, the process of recognition begins by comparing viewer-centered and object-centered representations to stored structural descriptions of known objects (so-called object recognition units [ORUs]). The ORU acts as an interface between visual representations (which describe what an object looks like) and semantic information (which describes the object's functional properties and attributes). According to the model, when information in viewer- and object-centered representations adequately matches structural information in some ORU, the ORU becomes activated. This, in turn, gives rise to a sense of familiarity and unlocks semantic information about the object. Since the ORU receives independent input from viewer- and object-centered representations, it can be activated by either independently if a sufficient match is obtained. Name retrieval occurs in the final stage of the model. Its position at the bottom of the model assumes that the semantic system does not contain a
AGNOSIA

Identification is immediate and certain when the patient is presented the stimulus in other sensory modalities.

When the patient fails to name but can indicate visual recognition by verbal description or gesture, the failure is usually considered to be anomic in nature. Unlike the agnostic, the anomic generally does not improve when the material is presented through another sensory modality (Spreen et al., 1986; Goodglass et al., 1968), and he is less apt to perform normally when asked to produce lists of words in specific categories, to complete open-ended sentences, or to respond to definitions. The conversational speech of the anomic may alert the examiner to the possibility of difficulty in visual confrontation naming because it contains word-finding pauses, circumlocutions, semantic paraphasias, and a general lack of substantives (see Chapter 2). We will later consider a syndrome called "optic aphasia," in which the naming disturbance is disproportionately severe for visually presented objects, and will consider whether this syndrome is anomic or anomic.

Visual agnosia has been classified in a number of different ways. The most widely known classification is Lissauer's (1970) apperceptive-associative distinction discussed earlier, which, from a clinical perspective, is based primarily on the success of the impairment. Visual agnosia has also been classified according to the specific category of visual material that cannot be recognized. Impairments in the recognition of faces (prosopagnosia), colors (color agnosia), objects (object agnosia), and an agnostic inability to read (agnosia alexia) have been described in isolation and in various combinations (Farah, 1991). The co-occurrence of associative visual object agnosia with alexia, color agnosia, and prosopagnosia is common, though not obligatory.

Some types of agnosia (e.g., visual object agnosia) involve a defect that prevents the recognition not only of the specific identity of an object but also of the general semantic class to which it belongs. Other forms of agnosia (e.g., prosopagnosia) are characterized by an ability to recognize the general nature of the object (e.g., a face), but a profound inability to appreciate its individual identity within that class. It remains to be determined whether the distinction between agnosia for object classes and agnosia for specific identities reflects a feature of brain organization or whether it represents the fact that visual and semantic information play different roles in the recognition of specific classes of objects (see below). It is possible that specific identity discriminations (e.g., "that's my wallet"), are more visually or semantically demanding than more general ones (e.g., "a wallet") and that certain classes of objects (e.g., places) place special demands on sensory-perceptual systems (Damasio et al., 1982; Warrington and Shallice, 1984). Alternatively, it is possible that certain classes of objects demand qualitatively different types of processing that differ in kind, rather than degree, from processes involved in recognizing other objects (Farah et al., 1990). We will consider these issues later when we describe some strikingly specific agnosias.

Apperceptive Visual Agnosia

The term apperceptive agnosia has been applied to a broad spectrum of patients who have in common some measurable impairment at the perceptual level, but whose elementary sensory functions appear to be relatively intact. Farah (1990) points out that the term has been applied to a heterogeneous range of disabilities from patients whose visual impairments prevent them from negotiating their surroundings (Luria et al., 1965) to those without in vivo impairments in object recognition who fail specialized perceptual tests that require patients to match stimuli across different views (Warrington and Taylor, 1973). Most cases have been associated with pathological processes such as carbon monoxide poisoning (von Hagen, 1941; Adler, 1944; Benson and Greenberg, 1969; Mendez, 1988; Milner and Heywood, 1988; Milner et al., 1991; Spar et al., 1991), mercury intoxification (Landia et al., 1981), cardiac arrest (Brown [case II], 1975), bilateral cerebrovascular accident (CVA) (Stauffenberg, 1914), basilar artery occlusion (Caplan, 1950), or bilateral posterior cortical atrophy (Benson et al., 1988; Mendez et al., 1990). The behavior of these patients suggests severe visual difficulties. Many are recovering from a state of cortical blindness. Because of their helpfulness in the visual environment, many are considered blind until they report
that they can indeed see, but not clearly or un-
til they are observed avoiding obstacles in their
environment. Standard testing then reveals
normal or near-normal acuity in the spared vis-
ual field. Preservation of sufficient field and
acuity to allow for recognition distinguishes the
apprehensive agnosia from the patient with An-
ton's syndrome (Anton, 1890), denial of corti-
cal blindness.

Apprehensive agnosia full recognition tasks
were defective in perceptual processing.
They cannot draw misidentified items or match
them to samples. They are generally unable to
point to objects named by the examiner. The
impairment most often involves elements of
the visual environment that require shape and
pattern perception (faces, objects, letters). The
recognition of even the simplest of line draw-
ings may be impossible. However, bright and
highly saturated colors may be better recog-
nized. Some patients can trace the outines of
letters, objects, or drawings (Goldstein and
Gelb, 1918; Landis et al., 1983), but often re-
trace them over and over because they lose the
starting point. Many patients behave as if they
are unaware or unconcerned about their deficit
until they are given a visual recognition task.
They will then acknowledge that they do not see
clearly. Others are aware of their difficulty
but try to conceal it.

Many patients complain that their visual en-
vironment changes or disappears as they try to
scrutinize it. Recognition may improve when
visual stimuli are moved or, in the case of read-
ing, if letters are traced (Botez, 1975). This
condition, known as visual static agnosia, has
been thought to reflect residual capacity of the
subcortical (extrapolar) visual system (Denny-Brown and Fischer, 1976; Zihl and
Von Cramon, 1979; Celesia et al., 1980). Pa-
ients may claim they need new glasses, or may
complain about poor lighting or the fact that
they have not had much prior experience with
the particular kind of visual material that they are
being asked to identify. One of our patients,
a retired architect, condescendingly remarked
about the poor quality of our line-drawn stim-
uli. It has always been difficult to characterize
the visual performance of these patients be-
cause of large inter- and intrasubjindvidual
variability, and in fact there may be multiple forms
of deficit (Shelton et al., 1994). Cognizant of
this variability, Farah (1990) attempted to
bring order out of chaos by subdividing appre-
ceptive agnosia into the following four behav-
iorally meaningful categories.

Narrow Apprehensive Agnosia. Representative
cases in this category include patients reported
by Adler (1944), Alexander and Albert (1983),
Benson and Greenberg (1980), Campan and
Latto (1985), Goldstein and Gelb (1918), Lan-
dis et al., (1982), and Milner et al. (1991). These
patients all have seemingly adequate ele-
mental visual function (acuity, visual fields,
lumiance detection, color vision, and depth
and movement perception) but display a strik-
ing inability to recognize, match, copy, or dis-
 criminate simple visual forms. Benson and
Greenberg's patient is a case in point.

The patient was a 25-year-old victim of ac-
cidental carbon monoxide poisoning. For sev-
eral months he was thought to be blind and yet
was seen one day navigating the corridor in his
wheelchair. He could often follow moving visual stimului, yet could not iden-
tify by vision alone objects placed be-
fore him. He could occasionally identify the
letters s and o if allowed to see them drawn or
if they were moved slowly before his eyes. Visual
acuity was at least 20/100, measured by his abil-
ity to indicate the orientation of the letter E,
detect the movement of small objects at stan-
dard distances, and reach for fine threads on a
piece of paper. Optokinetic nystagmus was
elicited bilaterally with fine 1/8 inch marks on
a tape. Visual fields were normal 3 mm-wide
objects with minimal inferior constriction bi-
laterally to 3 mm red and green objects. There
was an impairment of gaze with quasi-
random searching movements, particularly
when inspecting an object. His recognition
deficit included objects, photographs, body
parts, letters, and numbers, but not colors. He
could tell which of two objects was larger and
could detect small movements of small
targets. He easily identified and named objects
when he could touch them or hear the sounds
they made. He guessed at the names of objects,
utilizing color, size, and reflectance cues. He
was totally unable to match or copy material
that he could not identify. However, he was
taught to apply a name to each object in a small
agnosia. The term "simul-
taneous" was coined by Weigert (1994) to refer
to a condition in which the patient is unable
to appreciate the meaning of a whole picture
or scene even though the individual
parts are well recognized. Luria (1989) used
the term literally to indicate the inability to see
or attend to more than one object at a time.
Luria's use of the term has more generality
since it is debatable, even in mildly impaired
cases, whether the parts are recognized nor-
man.

Representative cases have been re-
ported by Hécaen and de AjuriaGuevara
(Hécaen, 1984; Holmes, 1918; Holmes and Horrax, 1919,
Luria (1989); Luria et al., (1965); and Tyler
(1968). Most of these patients sustained bilateral
parieto-occipital damage, although cases with
superior occipital (Rizzo and Hurtig, 1987) or inferior parietal damage (Kase et al.,
1977, case 1) have been reported. Simultaneg-
ognosia can also be present in the context of
generalized or localized degenerative disease
(Graft-Radford et al., 1993; Ardila et al., 1997;
Bewersdorf and Heilman, 1998; Mendez and
Charrier, 1998; Mendez, 2000).

As a result of their visual deficit, these pa-
tients are impaired in counting tasks (Holmes,
1918) and on tasks that require the naming of a
number of objects presented together (Luria et al.,
1963). The disorder may also be evident in
a dramatic inability to interpret the overall
meaning of a line drawing or picture, with
performance on such tasks often being a hapha-
azard, inferred reconstruction of fragmented pic-
ture elements. On the basis of the study of such
patients, Luria (Luria, 1959; Luria et al., 1963)
concluded that simultagnosia represents a complex perceptuomotor breakdown of the
active, serial, feature-by-feature analysis neces-
sary for processing elements of a visual scene
or pattern. In the most severe cases, prominent
features available in the stimulus array may
themselves be fragmented and distorted.

Luria equates simultagnosia with a per-
ceptual defect often found as part of Balint's
syndrome (Balint, 1909; Husain and Stein,
1988); which consists of (1) psychic paralysis of
fixation with an inability to voluntarily look
into the peripheral field (Tyler, 1968; Lopes et al.,
1979), (2) optic ataxia, manifested by clumsi-
ness or inability to manually respond to visual

Dorsal Simultagnosia. The term "simul-
taneous" was coined by Weigert (1994) to refer
to a condition in which the patient is unable
stimuli, with mislocation in space when point- ing to visual targets (Holmes, 1918; Boller et al., 1975; Hasenm and Kupfer, 1975; Levine et al., 1976; Damsato and Benton, 1979), and (3) a disturbance of visual attention mainly affecting the periphery of the visual field and resulting in a dynamic concentric narrowing of the effective field (Hécaen and Ajurriaguirre, 1954; Levine and Calvano, 1975; DeRenzi, 1982). Balint’s syndrome is almost invariably associated with large biparietal lesions (but see Watson and Rapcsak, 1987), and is especially severe when fronto-limbic lesions are also found (Hécaen and Ajurriaguirre, 1954). Frontal lobe involvement may lead to particularly severe psychic paralysis and optic ataxia, presumably because of disruption in visual-motor mechanisms and because of the role played by frontal eye fields and surrounding prefrontal cortex in the control of saccadic eye movements and visual attention (Lynch and McClaren, 1989).

Visual fields may be normal by standard perimetric testing but shrink to “shallow vision” when the patient concentrates on the visual environment. Performance may be worse in one hemifield, more often on the left. A striking example of narrowing of the “effective visual field” is given by Hécaen and Ajurriaguirre (1954, case 1). While their patient’s attention was focused on the tip of a cigarette held between his lips, he failed to see a match flame offered him and held a few inches away. A good example of this deficit is presented by Tyler (1968), whose patient suddenly developed visual difficulties after segmental basal artery occlusion.

Visual acuity was 20/20 with glasses. Visual fields were at first considered normal, but careful retes- testing showed that while the left field was normal to movement of large objects, these objects faded from awareness in one or two seconds. With continued testing within that field, awareness of even the movement of large objects was lost. In the right visual field, the central two degrees around fixation were always normal, the surrounding outer 20 degrees fatigued rapidly, and beyond 20 degrees, movement was recognized but objects faded rapidly. The patient could see only one object or part of one object at a time with his center two to four degrees of vision. She scanned normally when looking at predictable objects such as a circle or a square but frequently lost her place when viewing objects and pictures. Slight movement of her head made her lose her place. She reported seeing bits and fragments. For instance, when shown a picture of a flag, she said, “I see a lot of lines, now I see some stars.” When shown a dollar bill, she saw a picture of George Washington. Moments later when shown a cup, she said, “A cup with a picture of Washington on it.” Eye movement studies revealed a normal number of visual fixations per unit of time and a normal pattern of fixation for small saccades or so-called visual steps. However, there were very few, if any, so-called long saccades or leaps that relate one part of the picture to another.

The three subjects described by Rizzo and Hurtig (1987) reported intermittent disappearance of a light target during electrooculogram (EOG)-verified fixation. Rizzo and Hurtig argue that a disorder of attentional mechanisms that permit sustained awareness of targets is involved. Verfaellie et al. (1990), using Posner’s attentional cueing task, found that their Balint’s patient had difficulty shifting attention to the left or right visual field, and benefited only from cues directing attention to the upper visual field. This patient also demonstrated spontaneous blinking, which may normally participate in a complex system mediating saccadic eye movement, sensory relay, and attentional deployment (Watson and Rapcsak, 1987).
adaptation time are not, even in the presence of dementia, sufficient in themselves to produce an agnosia-like recognition defect. It is true, however, that many patients with visual agnosia have elements of this type of disturbance and many also have abnormalities in visual attention, search, and exploration. Recent work suggests that there may be heterogeneous deficits within the general category of apperceptive agnosia. Studies in which stimulus manipulations seek to mimic apperceptive agnosia in normal subjects point to the special importance of perceptual grouping processes in producing the defect (Vecera and Gilda, 1988). In some cases, a combination of impaired feature recognition and limited cognitive resources available for processing demanding visual material may be present (Grossman et al., 1997). It seems likely that such defects represent a continuum on which impairment is a necessary, but not sufficient, characteristic of agnosia.

Although each of the four types of apperceptive agnosia has a relatively consistent lesion profile, it is impossible with our present level of knowledge to fully specify the functional anatomy underlying the various forms of visual apperceptive agnosia. It is clear that the specificity called "apperceptive visual agnosia." Behaviorally, these patients differ along attentional, perceptual, oculomotor, and mnemonic dimensions; the relative contribution of each of these factors remains to be fully elucidated. However, it seems reasonable on clinical and experimental grounds to conclude that complex visual abilities are made up of dissociable ("modular") information-processing streams, including form discrimination, color perception, luminance, size, movement, and spatial localization and integration (Sprague et al., 1977; Pererin and Jeannerod, 1975; Berlyk and Sprague, 1975; Maunsell and Newcombe, 1987; DeYoe and Van Essen, 1988) and that the variability among apperceptive agnosics reflects the fact that these streams can be impaired singly or in combination at the individual case level.

Associtative Visual Agnosia

The major distinguishing feature of associative visual agnosia is the presence of a modality-specific object identification defect in the context of a preserved ability to copy and/or match stimuli presented in the affected modality. Preserved copying or matching has often been taken as evidence of "normal" perception, an assumption that has been called into question by recent evidence (see below). Although, as we shall see, perception is not entirely normal in these patients, the degree of perceptual disturbance in this class of patients is different in degree and kind from that seen in apperceptive agnosia. A number of well-documented cases meeting the above criteria have appeared in the literature, leaving no doubt about the existence of this form of agnosia (Rubens and Benson, 1971; Taylor and Warrington, 1971; Lhermitte et al., 1973; Benson et al., 1974; Hécaen et al., 1974; Newcombe and Ratcliff, 1974; Albert et al., 1975a, 1975b; Boller and Ratcliff, 1977; Pillon et al., 1987a; Davidoff and Wilson, 1985; McCarthy and Warrington, 1986; McCarthy and Warrington, 1986; Reddick and Humphreys, 1987a; Feinberg et al., 1994).

Pointing to objects named by the examiner, while characteristically impaired, may be better than identifying objects verbally or by gesture. This dependency on locally available transformation takes place in the context of a closed set, while naming an object involves an almost infinite list of potential names. It is also possible that pointing to a named object is an easier task because it does not involve speaking and therefore reduces the chances that an incorrect verbal response will be substituted for the correct one. In the case of stimulus recognition, in many patients, picture identification is more impaired than the identification of real objects and identification of line drawings is more impaired than either of these. A disturbance in the identification of line drawings or pictures may be the only residual defect after the acute disturbance has cleared. This dissociation is not seen in the naming performance of aphasics (Corlew and Nation, 1975; Hasfield and Howard, 1977), and may serve as a marker for the presence of agnosia in naming tasks.

Impairment of object recognition of faces (prosopagnosia), color (so-called color agnosia), and of written material (alexia) are frequently but not invariably found with associative object agnosia (Farah, 1991). Object agnosia itself is more rare than these other conditions, each of which may occur in isolation or in various combinations. The patients of Hécaen and Ajuriguerra (1956) and Lhermitte and Beauvois (1973) had no impairment in facial recognition, and reading was spared in the patients of Davidsen (1956), Newcombe and Ratcliff (1974), Mack and Boller (1977), and Albert et al. (1975a). Levine's patient and case 1 of Newcombe and Ratcliff had no color agnosia or alexia. Alexia is commonly found alone or with color agnosia (see Geschwind and Fussillo, 1968); prosopagnosia is sometimes an isolated recognition disturbance (De Renzi, 1986a; Fallis, 1955), but is often associated with achromatopsia (Critchley, 1965). Much debate has centered around the coexistence of these various signs. Some authors believe that symptom co-occurrence is a simple "neighborhood sign," while others believe it reflects the fact that certain classes of objects (e.g., faces and faces) overlap in their visual processing demands. Tactile and auditory recognition are typically intact, although two patients of Newcombe and Ratcliff (1974) and the patients of Taylor and Warrington (1971) and Feinberg et al. (1986) were unable to identify objects by touch or vision.

The associative agnostic not only cannot name seen objects but also typically fails when asked to demonstrate semantic knowledge about the stimulus or its functional properties. The failure to sort objects and pictures into categories will be markedly amnestic even if the subject can name objects, and he may be unable to identify the object at a more specific, individual level ("John's face" or "my wallet"). Others can identify neither the general class nor the individual within-class. Such differences may signal important differences among gnosis syndromes or may more simply reflect the fact that different recognition tasks demand different levels of specificity. For example, a "key" is often an adequate answer in an object recognition paradigm, while face recognition tasks require the subject to ascertain individual identity.

The role played by perceptual factors in associative agnosia has recently received attention. On the one hand, these patients are capable of remarkable visual achievements given the severity of their object recognition disturbances. The patients of Rubens and Benson, Taylor and Warrington, and Newcombe and Ratcliff (case 1) matched to sample and produced strikingly accurate drawings of pictures and objects they could not identify (Fig. 12-2). The patients of Rubens and Benson, and Taylor and Warrington were able to find hidden figures in figure-ground tests. Case 1 of Newcombe and Ratcliff showed no deficits on psychophysical tests of visual function.

On the other hand, two lines of evidence have made it clear that perceptual abilities are not normal in the vast majority of these patients and that such abnormalities may play a more active role in at least some aspects of the recognition disorder. First, although these patients may be capable of earning normal scores on tests of copying and matching, qualitative data suggest remarkably consistent evidence of slow, feature-by-feature, or "slavish" drawing that represents a reduced sensitivity to the more global aspects of the stimuli (Levine, 1975; Humphreys and Riddoch, 1987; Levine and Colavino, 1989; Farah, 1990; Suruki et al., 1997). For example, Humphreys and Riddoch (1987) provide an exquisitely detailed report of a patient, H.J.A., who took hours to complete remarkably accurate drawings of objects he could not identify. Our own experience with the prosopagnostic patient L.F. revealed that performance on difficult facial matching tests (e.g., Benton Test of Facial Recognition), though quantitatively normal, proceeded in a feature-by-feature manner, resulting in extremely prolonged response times. Second, systematic variation of perceptual variables can significantly affect the recognizability of stimuli (Levine and Colavino, 1978, 1989; Riddoch and Humphreys, 1987a). Stimulus complexity (e.g., presence of color, morphological similarity between items) appears to exert a strong effect on the frequency of semantic and morphological errors on object-naming tasks. Both the presence of fine-grained visual
information as well as the presence in the stimulus of “compound” (multiple) information appear to contribute to ease of identification (see Levine and Calvano, 1976). A frequently reported finding is that associative agnosia appears deficient in “gestalt perception,” although they may perceive local details relatively normally. Partially covering an item or placing it in an unusual context hinders identification. Levine and Calvano (1989) reported that their associative agnostic, L.H., was severely impaired on tasks of “visual closure”—the ability to perceive shape and identity of an object that has been degraded by visual noise (see also Farah, 1990). H.J.A., the patient reported by Humphreys and Riddoch (1987), performed poorly on a feature integration task requiring him to detect an upside down T among a group of upright Ts. Unlike normals, H.J.A. did not show faster detection when the stimuli were arranged in a discrete circular configuration (as opposed to random presentation), suggesting a deficit in integrating local features into an overall gestalt. These findings illustrate why there is growing discontent with the apperceptive-associative distinction, at least in its “strong” form. They also illustrate why parallel-distributed models of object recognition that posit no fundamental distinction between perception and memory/association (Goldman-Rakic, 1988; Damasio, 1989) deserve increased attention by neuropsychologists and behavioral neurologists interested in agnostic phenomena.

The cases of Kertesz (1979) and Wapner et al. (1978) further complicate the apperceptive-associative distinction. Kertesz’s patient presents a challenge, as she had elements of Balint’s syndrome, visual staticagnosia (Botes and Serbanescu, 1967; Botes, 1975), alexia without agraphia, prosopagnosia, and amnestic syndrome. The patient performed poorly on copying tasks (her reproductions were poorly executed and contained only fragmented elements of the associated target stimuli), but matched real objects; line drawings, colors, letters, and geometric figures better than she named or pointed to them. Verbal responses were marked by perseverations and form conflation. The patient had 20/20 acuity (open field method), and a spiraling visual field defect. A computed tomography (CT) scan revealed right frontal and deep left occipital lobe lesions.

Wapner and colleagues (1978) presented a case report of visual agnosia in an artist whose drawing skills were specifically assessed. Their patient suffered a cerebral infarction resulting in variable right hemianopia, visual recognition defect, and amnesia. A brain scan revealed bilateral medial occipital infarctions. Visual acuity was 20/70. The patient showed poor visual recognition of objects and drawings in the context of moderately impaired design copying. Interestingly, the patient showed a striking dissociation in qualitative drawing performance between objects he could and could not recognize. With unrecognized objects, his drawings revealed piecemeal, slavish reproduction of recognized elements. Describing his drawing of a telephone dial, he said, “a circle, another circle, a square . . . things keep coming out . . . and this is as though it books into something.” In contrast, when drawing an object he could identify, the patient relied on preserved structural knowledge of the essential components of the object, producing a sketch that was highly similar to the specific target as well as to the general class of objects to which the target belonged. He remarked, “can’t help but use your natural knowledge in drawing the thing.” These two cases are important for two reasons. First, they both showed dissociations among various tests classically used to tap the apperceptive level. Second, their combined deficits at the levels of perception and recognition underscore the fact that agnosia is the final outcome of many different defects, and again illustrate that the apperceptive-associative distinction should be viewed as an heuristic and perhaps nothing more.

In addition to the contribution of perceptual factors, there is evidence that the initial verbalized response to visual presentation can adversely affect recognition ability in at least some of these patients. Identification errors are usually morphological confusions or perseverations; though semantic errors are not uncommon. The tendency to perseverate previous naming errors and the disruptive influence of visual naming on tactile identification are examples of this. One might expect that visual agnosia patients with normal blindfolded tactile naming would perform at least as well when they simultaneously inspect and handle an object. However, the otherwise superior tactile identification of the patients of Ettinger and Wyke (1961) and Rubens et al. (1978, cited in Rubens, 1979) fell to the much lower level of visual identification alone when the patients were allowed to simultaneously view and handle the objects. Ettinger and Wyke’s patient, when given two exposures of each of 21 items, made 36 errors with vision alone, only 9 with touch alone, but 18 errors with vision and touch.

Prevention of the contaminating effect of verbal responses is not always easy. Many patients insist on speaking despite strict instructions to remain silent. Case 1 of Oxbury et al. (1969) was specifically instructed to demonstrate in silence the use of objects shown to her, but continued to name them aloud and then to produce an incorrect gesture that corresponded to her verbal misidentification. This same patient, when asked to match a line drawing to one of three real objects, would misname the drawing and then search in vain for the object corresponding to her incorrect name. It has been claimed that perseverations represent verbal reports of a lingering visual sensory ex
experience of previously viewed material (Crickley, 1964; Cummings et al., 1982). However, when patients are asked to draw an object on which such perseverative errors occur, they draw the item they are viewing, not the item whose name has been perseverated (Rubens and Benson, 1971; Lhermitte and Beauvois, 1973; Rubens et al., 1978, cited in Rubens, 1979). Successfully copying a misidentified picture generally does not facilitate identification of that picture. This suggests that the motor system generally does not have the ability to cue the visual identification process in most patients, although requiring the patient to write the name (Lhermitte and Beauvois, 1973) or to supply a description (Newcombe and Ratcliff, 1974) of an object instead of naming it aloud normalized recognition in one case and enhanced it in the other.

The most common visual field defect in associative agnosia is a dense, right homonymous hemianopia. In the patient of Albert and colleagues (1975a), the right visual field defect was confined to the upper quadrants. Two left-handed patients with left homonymous hemianopia have been reported (Newcombe and Ratcliff, 1974 [case 3]; Levine, 1978). Interestingly, reading was spared in all three of these patients. Normal visual fields have also been reported (Davidenko, 1956; Tayler and Warrington, 1971; Newcombe and Ratcliff, 1974 [case 1]).

The marked variability in performance of patients in the natural setting as opposed to the test setting was noted by Geschwind (1965), who viewed misidentification as a confounded response elaborated by the intact speech area pathologically disconnected from intact visual sensory area. Failure to supply the correct gesture results from concomitant disconnection between motor and sensory areas. The common association of visual object agnosia with right homonymous hemianopsia, alexia, and color agnosia, a triad occurring in the context of damage to mesial left occipital lobe and nearby posterior callosal fibers, supports the visual-verbal disconnection hypothesis. Authors arguing against the disconnection hypothesis cite (1) the occasional finding of normal visual fields or left homonymous hemianopia (Camber et al., 1980); (2) the occasional absence of color agnosia and alexia in the same patient (Newcombe and Ratcliff, 1974 [case 1]; Levine, 1978); and (3) the question of whether a left occipital-splenial lesion produces the syndrome of alexia without agraphia commonly but that of object agnosia only rarely. The pathology in some cases is inconsistent with a visual-verbal disconnection view. For example, Levine's (1978) patient had a unilateral right occipital lobe resection and was, in fact, able to verbally code some visually presented stimuli with remarkable accuracy (e.g., "something with "U" in it," when looking at a padlock). In a series of recent cases of associative visual agnosia, consistent damage to the dominant parieto-occipital, fusiform, and lingual gyr, but inconsistent damage to the callosal splenium was found (Feinberg et al., 1994).

Strictly speaking, the imperfect correlation between color agnosia, alexia, and visual object agnosia does not, by itself, invalidate the visual-verbal disconnection hypothesis. It remains possible that there may be highly specific forms of visual-verbal disconnection, and that unilateral or bilateral intrahemispheric disconnection and/or selective destruction of independent pathways mediating various elements of visual recognition plays a role (see Ratcliff and Ross, 1981). There is, for example, evidence for the specificity of neural pathways for color (Zeki, 1973, 1977; Meadows, 1974a). This kind of specificity is also implicit in the classical work of Hubel and Wiesel (1977). However, more damaging for the visual-verbal disconnection view is the fact that many associative agnosics fail on tasks that require such knowledge, such as functional classification or gesturing.

Neuropsychological data in associative agnosia present a confusing picture. Some authors (Warrington, 1985) suggest that diffuse damage may be involved, but that a lateralized left hemisphere lesion may be sufficient. Alexander and Albert (1983) argue for a bilateral occipitotemporal localization (see also Benson et al., 1974). Geschwind's (1965) visual-verbal disconnection model is based on a unilateral left mesial occipital localization, and other left-sided cases include the patients of Warrington (1971) and Feinberg et al. (1986, 1994). Levine's (1978) patient shows that associative agnosia can result from a unilateral right occipital lesion. Farah (1990) suggests that this heterogeneity might account for different perceptual impairments in patients with associative agnosia. For example, in the context of the debate regarding the relative significance of cortical and white matter lesions (Albert et al., 1979; Ross, 1980b), it seems possible, even likely, that there are multiple forms of associative visual agnosia representing impairment at different levels of processing (Damasio and Damasio, 1982). Patients such as those of Taylor and Warrington (1971) and Newcombe and Ratcliff (1974 [cases 1 and 2]) with diffuse bilateral disease processes, tactile agnosia, and normal visual fields probably form a separate group from those with right homonymous hemianopia associated with infarction in the territory of the left posterior cerebral artery.

Optic Aphasia

The term "optic aphasia" was introduced by Freud (1880) to describe the deficit of one of his patients with a right homonymous hemianopia and agnosia due to a left parieto-occipital tumor; the patient's naming ability was impaired primarily for visually presented objects. The case report is of little value because of its incompleteness, but Freud's speculations are pertinent. He hypothesized that a left speech area-right occipital disconnection was the basis for the visual naming deficit in the intact visual field. In current usage, optic aphasia refers to the condition in which patients are unable to name visually presented objects but are able to show that they recognize the object by indicating its use, by pointing to it when it is named, or by otherwise demonstrating knowledge of object meaning. Tactile and auditory naming are preserved. Representative cases have been reported by Lhermitte and Beauvois (1973), Riddoch and Humphreys (1987b), Larabee et al. (1985), and Coslett and Saffran (1988b). Whether optic aphasia and associative agnosia differ in degree or kind remains a matter of controversy (De Renzi and Satz, 1977; Chanoine et al., 1998), though the recent trend is to consider it a separate entity.

The patient of Lhermitte and Beauvois (1973) represents a good example of the syndrome of optic aphasia. Their patient suffered a left posterior cerebral artery territory infarction and presented with right homonymous hemianopia, moderately severe amnesia, and alexia. The most striking feature of his presentation was an inability to name visually presented objects. Naming errors consisted of perseverations of previously presented objects, semantic errors (in which an object was given the name of a semantically related object), and, less frequently, visual errors. At the same time, he could demonstrate that he knew what the object was by demonstrating how it would be used. Drawing of viewed objects was normal even when the object was mismatched. He showed normal tactile naming in both right and left hands and could name environmental sounds with little difficulty. Also, when given the name of an object he was viewing, he could provide an accurate definition of the object and its functional properties. Although he was aware of his visual field defect and his reading disturbance, he was not aware of his difficulty in visual naming.

What makes optic aphasia so important is that it challenges the widely held view that object naming is based on a common set of semantic representations of known objects that can be accessed from any sensory modality. The problem is this: why should a patient who shows intact verbal semantics (as evidenced, for example, by his good performance on the definitions task) and intact visual semantics (as evidenced by his accurate naming of object use) be able to name only those objects that are held in the hand? There have been several different answers to this question. Beauvois (1982) suggested that, in J.F., visual and verbal semantics became disconnected from each other, and that only tactile input had preserved access to verbal semantics. This view is based on the more general idea of multiple, modality-specific semantic systems, segregated according to the modality in which constituent information is represented (Warrington, 1985; Shallice, 1988).

Riddoch and Humphreys (1987b) suggest a different answer, based on their analysis of another case of optic aphasia. Riddoch and Humphreys presented an object decision task in which the patient had to decide whether a
series of individual line drawings represented real objects or not. Their patient performed normally on this task, which suggests that he had preserved knowledge of object structure. However, the patient was impaired in grouping semantically related objects (e.g., hammer and nail), and could not draw named objects from memory. Thus, in either direction, the patient could not link semantic and visual information. On the basis of these results, Biddock and Humphreys describe optic aphasia as a modality-specific semantic access problem. They further postulate that the knowledge of how to use objects is linked to structural rather than to semantic properties, and suggest that the patient may be able to demonstrate object use by a direct connection between visual object recognition units and motor action systems.

A third answer was provided by Coslett and Saffran (1989a), who suggested that at least some patients with optic aphasia are able to access semantic information contained in the right hemisphere but are unable to access the semantic system and speech production mechanisms within the left hemisphere. Like Warrington (1985) and Shallice (1988), these authors posit multiple semantic systems, but argue that these systems are differentiated not by the modality of input but by their anatomic locus in the right or left hemisphere. The patient described by Coslett and Saffran was a 67-year-old man with a large left occipital lobe infarct and a lacunar infarct in the right internal capsule. The patient was unable to name visually presented objects but was able to name them to palpation and visual description and could point to them when they were named. This latter ability is not easily accommodated by a model that dissociates all visual and verbal input. The patient could correctly categorize unnamed pictures and words and could group unrecognizable objects according to their functional similarity and semantic association. This provides good evidence that the patient was able to access detailed semantic information from visually presented objects, which Coslett and Saffran argue reflects an intact right hemisphere semantic system.

While the mechanisms discussed above all invoke deficits of semantic access, Shuren et al. (1993) suggested that the critical deficit in optic aphasia may be lexical access. They reported three patients who named visually presented stimuli normally, but could not name the same objects when given a definition, nor could they demonstrate or describe the use of the objects. They called this disorder “non-optic aphasia” and suggested that while percepts could access an intact lexicon, the semantic representations were degraded. In contrast, patients with optic aphasia may be able to access semantics, but not the lexicon.

Although optic aphasia appears to be qualitatively different from associative agnosia, the possibility remains that, at least in some patients, the distinction may be a matter of degree (De Renzi et al., 1997; Chabot et al., 1998). Clinical lore suggests that patients may evolve into optic aphasia during recovery from classical associative visual agnosia. The fact that some patients can be made to oscillate between optic aphasia and visual agnosia by varying the instructions given them on a particular task illustrates, at least in some cases, the distinction between a naming disorder and a disturbance of recognition.

Color Agnosia

Patients with color agnosia are, by classic definition, unable to name colors shown to them or to point to a color named by the examiner, yet perform normally on nonverbal tasks of color perception. One of the earliest cases was reported by Wilbrand (1857), who referred to the defect as “amnesic color blindness.” Wilbrand observed that his patient could not find the appropriate word for a color displayed, that he frequently perseverated names across trials, and that naming the color of a familiar object out of sight was impaired. Wilbrand invoked an “amnesic” disorder because the patient often indicated that he had forgotten the name of the color he was shown.

The term color agnosia presents something of a conceptual dilemma. Unlike objects, colors cannot be heard or palpated and cannot be shown in use; they can only be known through vision or visual representation (imagination). It is difficult to imagine a clinical tool to assess color recognition in other modalities and thus it is hard to establish the modality specificity of the defect, although conceptual color tasks (e.g., “what color is associated with feelings of envy?”) have utility in assessing semantic aspects of color processing. Still, acquired anomalies of color vision and color performance do occur as a result of lesions to the posterior cortex (De Rensi and Spinler, 1987). Four syndromes of color disturbance have been described: (1) central achromatopsia/dyschromatopsia (MacKay and Dunlop, 1899; Green and Lessell, 1977; Meadows, 1974a; Pearlmutter et al., 1979; Damasio et al., 1986; Young and Fishman, 1990), (2) color anoma, which is found in association with pure alexia and right homonymous hemianopia and is attributable to visual–visual disconnection (Geschwind and Fuillo, 1966; Oxbury et al., 1969 [case 1]; Meadows, 1974a; Beauvois and Saillant, 1985); (3) specific color aphasia, in which the patient has linguistic defects but the impairment in utilizing color names is disproportionately severe (Kinbourne and Warrington, 1964; Oxbury et al., 1969 [case 2]); and (4) color-naming and color association defects concomitant with aphasia (De Rensi et al., 1972, 1973; Wyke and Holgate, 1973; Cohen and Keltner, 1978). We will review the first three of these defects below. A summary of performance defects in patients with these various syndromes is presented in Table 12–1 (Bowers, 1981).

Central Achromatopsia/Dyschromatopsia. Central achromatopsia refers to a loss of color vision due to central nervous system (CNS) disease. The causative lesions can be in the optic nerve, chiasm, or in one or both of the cerebral hemispheres (Green and Lessell, 1977). The disorder can be hemianopic (Albert et al., 1975b), or can exist throughout the visual

<p>| Table 12-1. Summary of Color Performance Deficits in Patients with Various Syndromes |</p>
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<td>Hue discrimination</td>
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<td>Verbal–verbal</td>
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<td>Naming items of specific colors</td>
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<td>Verbal–verbal</td>
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<td>Color–object pointing</td>
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The symbol + refers to intact performance and – refers to impaired performance.

*Performance depends upon severity of achromatopsia. In mild cases, only hue discrimination is impaired.

*Performance on these tasks is impaired as long as patient does not attempt to verbalize answers. If patient does attempt to do so, then verbalizations can interfere with performance.

*Global and Wernicke's aphasics are impaired; all other aphasics are OK.
lesions anterior to V4, but completely sparing it, can lead to achromatopsia in monkeys. Damasio et al. (1980) speculate that the lingual and fusiform gyr in humans may be the homologues of area V-4. The exact location is not currently known, though recent functional imaging evidence points to the fusiform gyrus (Sakai et al., 1998). What does seem clear from clinical data is that one single area in each hemisphere (centrally located in the lower visual association cortex) controls color processing for the entire hemifield (Damasio et al., 1980).

Color Anomia. The patient with color anomia succeeds on visual-verbal and verbal-verbal tasks, but is unable to name colors, a visual-verbal task. The disorder is usually associated with the syndrome of alexia without agraphia (Stengel, 1948; Geschwind and Fusillo, 1960; Carlesimo et al., 1998), and frequently exists in the context of right homonymous hemianopia. The underlying neuropsychological mechanism is a visual-verbal disconnection resulting from infarction in the left posterior cerebral artery (PCA) distribution. The patients of Geschwind and Fusillo (1960) and Oxbury et al. (1969 [case 1]) are classic examples of this syndrome.

These patients may show impairment on some tasks related to color perception, such as coloring pictures or detecting errors in wrongly colored stimuli. This impairment is exacerbated if patients attempt to "verbalize" their answers (Bowers, 1983). Damasio and colleagues (1979) suggest that the type of stimulus, the demands of the task, and the patient’s problem-solving approach can strongly influence the extent of visual-verbal dissociation. In their analysis, visual-verbal dissociation is maximized when, at the perceptual level, stimuli are purely visual (such as color), structurally less rich, or low in associative value. At the verbal end, visual-verbal dissociation is maximized when a specific name, rather than the name of a broad category, is involved (Boller and Spinnler, 1967). If the patient’s verbalizations about the stimulus are incorrect, they may interfere with attempts to assign it the correct color.

AGNOSIA

Specific Color Aphasias. Patients with this syndrome are distinguished from color anoma by their poor performance on verbal-verbal tasks. The patients of Oxbury et al. (1969 [case 2] and Kinsbourne and Warrington (1964) are among the best documented cases of this variety. Aphasic symptoms are usually present, but the difficulty with color names and other color-associated skills is disproportionately severe. The patient of Oxbury et al. had head trauma (and probable bilateral lesions) with complete right homonymous hemianopia and mild right hemiparesis. Kinsbourne and Warrington’s patient had a left posterior parietal subcortical hemiatoma. These patients can generally sort colors categorically and according to hue, and can appropriately match colors. These deficits are similar to those reported in aphasic patients by De Renzi et al. (1972).

Prosopagnosia

The term "prosopagnosia" was formally introduced by Bodamer (1947) to describe an acquired inability to recognize previously familiar faces. Patients with this disorder often present with a dramatic and recognizable disability that comes to the attention of the clinician by personal encounter or by the report of distressed family members who are concerned about the patient’s inability to recognize them. Typically, an inability to recognize newly encountered individuals (e.g., health-care workers and hospital personnel) is also present. The inability to recognize family members, friends, and hospital staff may lead to the mistaken conclusion that the patient is suffering from a severe memory defect or a generalized dementia. The disorder should be distinguished from Capgras syndrome, a psychiatric disturbance in which the patient believes that familiar persons have been replaced by imposters (Syndouin et al., 1978; Alexander et al., 1978; Shabab and Weitzen, 1979). When formally tested, patients with prosopagnosia may score normally on face discrimination and matching tasks (Warrington and James, 1967; Benton and Van Allen, 1972). They almost invariably recognize faces as faces, and can often succeed at perceptually demanding tasks of age, gender, and expression discrimination (Tzavaras et al., 1970; Benton and Van Allen, 1972). Their primary deficit is in identifying and recognizing whose face they are viewing. Such patients may be unable to recognize their own face in a mirror, though they often correctly infer that it must be themselves. The defect prevents the identification of famous personalities, family members, and other familiar persons, and often severely limits the ability to acquire familiarity with faces first encountered after illness onset (Beyn and Krayneze, 1968).

Prosopagnosia typically presents as an acquired disorder, although convincing cases in which the problem in recognition exists on a developmental basis have been reported (De Haan et al., 1991; De Haan and Campbell, 1991; Krecke, 1994; Ariel and Sadle, 1996; Duchaine, 2000). The patient typically reports, and demonstrates in everyday behavior, an inability to recognize the identity of individuals by their facial features. However, the patient is easily capable of recognizing them by voice or some other nonfacial cue. Prosopagnosia typically presents as a disabling condition, but affected patients are often able to compensate remarkably after an initial period of adjustment. For example, such patients often become able to use extrafacial cues, including clothing, character, gait, length of hair, beard, or distinguishing birthmark, to achieve recognition, though significant individual differences exist. In many cases, patients realize that they have a defect and are distressed by it, although there are cases in which the patient is unaware of the problem (Bodamer, 1947). The variability in clinical presentation suggests that the clinician should always carefully evaluate the scope of the defect as well as deficit awareness. Clinical examination often reveals additional neuropsychological deficits including central achromatopsia, constructional difficulty, topographical memory loss, and dressing apraxia, and the patient may or may not show more generalized signs of object agnosia (Hécaen and Angelergues, 1963). As with many cases of recognition disturbance, there is evidence that prosopagnosia may take apperceptive and associative forms (Gleason et al., 1970; Levine, 1978; Bauer and Trobe, 1984; De Renzi et al., 1972).
cases may appear with unilateral (right hemisphere) damage, much like the occasional case of "crossed aphasia" (Aptman et al., 1977). Some authors have distinguished among different types of prosopagnosia, each with its particular lesion profile. Damasio et al. (1990) described three subtypes: an associative type characterized by bilateral damage in Brodmann area 18, 19, and 37; an amnesic-associative type resulting from damage to the hippocampus and surrounding cortex; and an apperceptive type resulting from extensive right hemisphere damage involving Brodmann areas 18, 19, and 37. Likewise, De Renzi et al. (1981) distinguish between apperceptive and associative prosopagnosia.

Superior field defects (either homonymous hemianopia or superior quadrantanopia) are most common, suggesting that causative lesions tend to occur inferiorly in the occipitotemporal region (Meadows, 1974b). Some authors believe that unilateral right hemisphere disorders are responsible for inferior occipitotemporal damage, which is consistent with the pattern of spared and impaired abilities in these patients that has led to the construction of models of the face recognition process. One critical question that still awaits final resolution is whether face recognition is specific to the particular class of faces or whether it is simply a matter of the brain's ability to recognize specific features. This question remains to be answered.

The occipitotemporal projection system (OPTS) is a series of short U-fibers through the cortical mantle to connect adjacent regions in striate, prestriate, and inferior temporal cortex (Tusa and Ungerleider, 1985). This system, which serves as the functional interface between visual association cortex and temporal lobe, has been particularly implicated in prosopagnosia (Benson et al., 1974; Meadows, 1974b; Bauer, 1982). The regions in temporal lobe served by the OPTS subsequently project to the limbic system. Accordingly, prosopagnosia has been interpreted as a visual-limbic disconnection syndrome (Benson et al., 1974; Bauer, 1982). Evidence that prosopagnosia suffer from reductions in emotional responsivity to visual stimuli seems to support this idea (Bauer, 1982; Habib, 1986). However, the situation seems more complex, because OPTS lesions anterior to the occipitotemporal area (which, if it occurs, would produce a visual-limbic disconnection) do not typically result in prosopagnosia (Meadows, 1974b). Intrinsic damage to the occipitotemporal area thus seems important. It is possible that intrinsic damage to the occipitotemporal region destroys or disconnects from visual input the association cortex in which visual representations of faces (or at least the "hardware" for activating such representations) reside (Damasio et al., 1985; Perrett et al., 1985; Perrett et al., 1982). Second, occipitotemporal lesions involving the OTHS are usually posterior enough to directly affect portions of inferotemporal cortex, a region that seems particularly critical in recognizing, categorizing, and discriminating visual forms (Iversen and Weiskrantz, 1964, 1967; Kuypers et al., 1965; Gross et al., 1972). This may contribute to the "underspecification" of visual detail, which recent authors (Lieb et al., 1978; Schleseworth et al., 1982) have considered important in the production of the defect.

Prosopagnosia has stimulated remarkable interest because it apparently represents a category-specific defect and thus may shed light on the manner in which perceptual representations are organized in the brain. The pattern of spared and impaired abilities in these patients has led to the construction of models of the face recognition process. One critical question that still awaits final resolution is whether face recognition is specific to the particular class of faces or whether it is simply a matter of the brain's ability to recognize specific features. This question remains to be answered.

An important component of the model is the "face recognition unit" (FRU), which receives input from structural encoding, and contains the visual structural descriptions that allow a particular known face to be discriminated from other familiar and unfamiliar faces. According to the model, each familiar face has its own FRU. Activation of the FRU leads to a sense of familiarity, but not to individual identification, since the FRU contains structural, but not semantic information. Activation of the FRU normally leads to the activation of a "person identity node" (PIN) that contains semantic information about the owner of the face. After activation of the FRU and PIN, a name might be assigned to the individual face. According to the model, naming occurs only after an appropriate PIN is activated.

This kind of model has been useful in organizing a large body of empirical findings on face recognition in normal subjects and is a useful.
heuristic in understanding patterns of impairment in face processing that occur after brain injury. As applied to prosopagnosia, the model is useful in describing the various stages at which a face recognition defect can occur. Some patients have difficulty at the structural encoding level, and thus have difficulty with a broad range of face discrimination and perception tasks. Others have deficits much later in the processing chain. In some patients, the face recognition units themselves appear to be damaged, while in others, access to otherwise intact FRUs, or from the FRU to the PIN, appears to be impaired. Evidence of intact FRUs comes from case studies in which patients more easily learn correct face-name matches than incorrect ones (Bueray, 1991; De Haan et al., 1991), and in studies demonstrating intact access to person information in semantic priming paradigms (Young et al., 1985).

Recent computational studies have attempted to use computer simulations to evaluate the degree to which impairments at particular levels of a cognitive model could account for the pattern of spared and impared face identification abilities in at least some prosopagnosics. Using an interactive activation model patterned after similar models in word recognition, Burton et al. (1990) attempted to simulate the behavior of their prosopagnosic patient, P.H. This patient failed at all conventional tests of facial recognition, but showed intact semantic priming of familiarity decisions with both faces and names. That is, when Prince Charles' face was presented as a prime, he was more likely to recognize Princess Diana's face as familiar than an unrelated face. The authors simulated the behavior of their prosopagnosic patient, P.H. This patient failed at all conventional tests of facial recognition, but showed intact semantic priming of familiarity decisions with both faces and names. That is, when Prince Charles' face was presented as a prime, he was more likely to recognize Princess Diana's face as familiar than an unrelated face. The authors attempted to simulate this finding by "lesioning" their interactive activation model. The basic architecture of the model, patterned after the Bruce and Young (1986) face recognition model, contained a number of distinct pools of units corresponding to names, faces (FRUs), persons (PINs), and semantic information. In their simulation, input was applied to an individual name or FRU, and activation of associated units was measured. The normal behavior of the model accurately simulated cross-domain (face-name) and within-domain (name-name) semantic priming. For example, when input was applied to the Prince Charles PIN, increased activation (priming) occurred at the Diana FRU. On the basis of their behavioral data, Burton et al. hypothesized that P.H.'s lesion damaged the links between FRUs and PINS. In the normal simulation described above, all connections were equally weighted. To simulate damaged links between the FRUs and the PINs, the weights assigned to these links were halved and model was run again. Results from the simulations showed (1) normal name-name priming (expected in prosopagnosia, since the disorder does not affect the patient's ability to recognize names) and (2) intact cross-domain (face-name) priming for names despite the fact that activation at the FRU never reached recognition threshold. Thus, this computational model successfully simulated both normal and disordered face recognition performance.

Other researchers have developed computational models to address the issue of whether the pattern of spared and impaired recognition abilities in prosopagnosia implies impaired access to an intact face recognition processor (Parah et al., 1983). Although the dominant view of covert recognition in prosopagnosia is that it represents a failure of the face processor to access conscious awareness, these researchers argued that the covert recognition phenomenon reflected damage to the face processor itself. They argued that the damaged neural network would manifest residual knowledge in the kinds of tasks used to measure face recognition. Studies with a three-layer model consisting of face, name, and semantic (occupational) information for 16 units. The model contained face input and name units on the first level, face- and name-hidden units on the second level, and semantic units on the third level. Using a contrastive Hebbian learning algorithm, the network was first trained to associate an individual's face, name, and semantics whenever one of these was presented. The network was then lesioned by removing between 2 and 14 units from the 16-teen face input pool and from the 16-teen face hidden unit pool. In four simulations measuring face-name learning, speed of visual percep-ception, semantic priming, and face recognition, they found that their lesioned model displayed behavior remarkably similar to that reported in experimental studies of prosopagnosia, showing covert without overt recognition.

Computational approaches are valuable because they provide dynamic, hypothesis-driven tests of models derived from clinical cases. Their success does not, of course, imply that the actual neural activity underlying face recognition is being accurately modeled. However, this overall approach does yield suggestive data regarding the structure of the neural architecture underlying face recognition abilities.

Recent neuroimaging studies have generally tended to confirm suspicions about functional location of face processing learned from patient studies, though it is apparent that many studies have found lateralized (left or right) rather than bilateral activations. Andressen et al. (1996) measured cerebral blood flow with positron emission tomography (PET) while subjects performed one of three face-processing tasks: classifying faces by gender, recognizing new faces, and recognizing familiar faces. Results showed activation of the left lingual gyrus and fusiform gyrus that was homologous to the left fusiform gyrus of normal subjects. However, the right hemisphere was also activated in the prosopagnosic patient. This is consistent with the lesion model in which the right hemisphere is responsible for processing novel faces.

There have been few electrophysiological investigations with prosopagnosic patients. In a recent study, unfamiliar faces, inverted faces, and house faces were shown to a prosopagnosic and a control subject. The controls showed consistent early negativity (N170) at the lateral temporal electrode sites, but the prosopagnosic did not (Eimer and McCarthy, 1999). This pattern was seen only for the faces. These data were interpreted as evidence of a deficit in structural encoding of faces in the prosopagnosic patient.

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Some electrophysiological data from human subjects is also relevant to the question of face processing. Studies with implanted electrodes have revealed patches of cells in extrastriate visual cortex that selectively respond to faces. Interestingly, evidence suggests that these cells are intermixed with others that are not face responsive, rather than forming a localized "face region" (Allison et al., 1994). In a study of facial repetition effects, scalp potential and current density maps revealed maximum activity in the inferotemporal and fusiform gyr (from 50 to at least 250 msec), mainly on the right, for both faces and shapes, and in the hippocampus and adjacent areas (around 300 msec), specifically for faces (George et al., 1997).

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ber of faces one must recognize, and familiar-
ity with specific exemplars, can influence recog-
nition in such a way as to make it appear
category-specific (Damasio et al., 1990).

One study (Farah et al., 1995a) questioned
this view by showing that a prosopagnosic
patient had more difficulty recognizing faces
than that in recognizing objects, even when the
two tasks were equated for difficulty. This
finding is relevant to the question of whether
faces are special, or whether they are some-
thing of an object category, and the fact that
many patients with complete visual-linguistic
disconnection (particularly those with an-
terior temporal lesions) or dienocplastic
pathology (e.g., alcoholic Korsakoff’s syndrome)
are not prosopagnosic, if one adopts the reason-
dable definition that a selective face recogni-
tion deficit in the retrograde compartment must
ever be present. In amnesic states, the impairment
in both anterior and retrograde memory is al-
most never absolute; such patients have a cli-
mically significant deficit in learning new infor-
mation and in retrieving from the remote store,
yet some new learning and remote memory is
variously possible and demonstrated by at least
some degree of judged confidence or familiar-
ity. In most forms of acquired amnesia, there is
a relative preservation of remote memory,
whereas in prosopagnosia, the recognition of
remotely learned faces (friends, family mem-
ers) is particularly impaired (Meadows, 1974b).
In prosopagnosia, the impairment in
face recognition is characteristically absolute
and inspection of faces yields no subjective fa-
miliarity with the viewed person. Finally, most
prosopagnosics, unlike amnesics, have signifi-
cant perceptual dysfunction, including impair-
ment in gestalt processing, and apparent de-
ficits in analyzing low spatial frequency
components of faces (Sergent and Vilenmue,
1989).

One prominent view is that the underlying
deficit prevents the identification of individual
items within a class of objects that are visually
similar (Damasio et al., 1982). According to
this view, recognition proceeds normally at the
superordinate level (i.e., the patient continues
correctly that she is looking at a face) but
breaks down at subordinate levels. Several rel-
levant factors, including the similarity in phys-
ical structure among facial exemplars, the num-

occipitotemporal regions and associated struc-
tures that are normally involved in initial per-
ceptual processing and encoding. Evidence
that the situation is, in fact, far more complex
is provided by a recent PET study (Haxby et
al., 1996) that evaluated neural substrates of
encoding and retrieval of new faces. A region
in the right hippocampus and adjacent cortex
was activated during memory encoding but not
during recognition. The most striking finding
in neocortex was the lateralization of prefrontal
participation. Encoding activated left pre-
frontal cortex, whereas recognition activated
right prefrontal cortex. These results suggest
that face recognition is not mediated simply by
recapitulation of operations performed at the
time of encoding but, rather, involves regions
that are anatomically distant from initial pro-
cessing zones.

The nature of the cognitive defect(s) under-
lying prosopagnosia has been a matter of sig-
nificant debate since the disorder was first for-
mally described. While it is obvious that no
single impairment can explain all the varieties
of prosopagnosia, it has been tempting to search
for a core deficit in cases in which ele-
mentary visual perception appears adequate to
the task of face recognition.

Some consider prosopagnosia to be one of
a number of category-specific agnosias. Although
in prosopagnosia the defect is identifying in-
dividuals on the basis of their facial features is
disproportionately impaired (compared, for ex-
ample, to the identification of objects), the bal-
ance of available data suggests that, in most
cases, impairment in recognizing other classes
of objects co-exists with the face agnosia. Doubt
is cast on a purely category-specific view of
reports of prosopagnosics who concurrently lost
the ability to recognize specific chairs (Faust,
1955) or automobiles (Lhermitte and Pillon,
1975). Bornstein and colleagues have reported
two prosopagnosics, one a birdwatcher, the other
a farmer, who concurrently lost the ability to
recognize individual birds or cows, respectively
(Bornstein and Kiddron, 1959; Bornstein, 1963;
Bornstein et al., 1969). Additional issues re-
garding the concept of category specificity are
discussed below.

Another view is that prosopagnosia repre-
sents a limited form of amnesia. This hypothe-
isis is supported by findings that patients with
prosopagnosia cannot learn and remember
new faces, because the causative lesions par-
tially or completely disconnect vision from lia-
ble structures important to initial encoding and
storage of information (Ross, 1980a; Bauer,
1982). The basic idea is that the structural rep-
resentations of faces that result from visual in-
spection cannot be matched to stored repre-
sentations built up from past experience.
However, doubt is cast on this line of reasoning
because it implies that many patients with comple-
tive visual-linguistic disconnection (particularly those with an-
terior temporal lesions) or dienocplastic pathol-
yogy (e.g., alcoholic Korsakoff’s syndrome) are not prosopagnosic, if one adopts the reasonable de-
inition that a selective face recognition de-
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ical structure among facial exemplars, the num-

ber of objects (Warrington and Shallice, 1984;
Warrington and McCarthy, 1987; Farah et al.,
1989; Damasio et al., 1990; Farah, 1991). As
indicated earlier, questions regarding category
specificity arise in the context of evaluating the
significance of other selectives forms of ag-
nosia such as prosopagnosia. Although some
prosopagnosics have a relatively pure face
recognition deficit (De Renzi, 1986a), others
have difficulty in recognizing objects in other
semantic classes, such as animals (Bornstein
and Kiddron, 1959; Damasio et al., 1982), plants
(Shaftelworth et al., 1982), or foods (Damasio
et al., 1982; Michel et al., 1986). When these
problems have been considered together, it has
been suggested that a supercategorical deficit
in recognizing living things might be involved
(see Farah et al., 1991), reflecting primary
neural organization along an animate-inani-
mate dimension (Caramazza and Shelton,
1998). However, it is unclear how other cate-
gory-specific deficits involving, for example,
musical instruments (Gainotti and Silvetti,
1986), medical implements, or other objects
(Shaftelworth et al., 1997), or cars (Ford et al., 1997)
can be accommodated in this framework (Dixon,
2000; Dixon et al., 2000).

Two general explanations for such category
specificity have been offered. One is that these
disorders represent localized impairment in a
semantic system that is organized according to
conceptual categories. Another view suggests
that living things are more visually complex
(Gloning et al., 1970), are more visually simi-
lar to each other (Ford et al., 1997), or re-
quire for identification more specific names
than do nonliving things (see Farah et al.,
1991). The former explanation suggests that a
selective disruption in the recognition of living
things reveals something basic about the struc-
ture of semantic memory; the latter implies
that such selective disruption results from task,
processing, or response factors that have been
confounded in the usual clinical tasks of object
recognition (Dixon, 2000).

With these alternative explanations in mind,
Farah et al. (1991) devised a series of object
naming tasks that took account of visual com-
plicity, interitem similarity, and response
specificity (e.g., “table” vs. “picnic table”) and
gave them to two patients who became agnostic
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mild, and do not prevent the patient from recog-
nizing with incoming information provided
audition is not required. Difficulties in temporal
auditory analysis and localization of sounds in
space are common. These disorders are rare,
and their underlying neuroanatomic basis is
poorly understood (Rosati et al., 1982). Some
case reports have questioned the distinctive na-
ture of "true" cortical deafness (Vignolo, 1969;
Lhermitte et al., 1971; Kanshelpsky et al.,
1973).

Distinguishing between cortical deafness and
auditory agnosia continues to be problematic.
It has been suggested that a diagnosis of
cortical deafness requires a demonstration that
brainstem auditory evoked responses are nor-
mal, but cortical evoked potentials are not
(Coletti et al., 1984). One distinction which is
frequently cited is that cortically deaf patients
feel deaf and seem to be so, whereas auditory
agnosics insist that they are not deaf (Michel
et al., 1980). This turns out to be a poor crite-
ron. Although it was originally believed that
cortical deafness evoked the primary
auditory cortex to total hearing loss, evidence
from animal experiments (Neff, 1961;
Massopust and Wolfa, 1967), cortical map-
ing of the auditory area (Celesia, 1976), and
clinical-pathological studies in humans (Wol-
fart et al., 1985; Mahoosdu et al., 1986) indi-
icate that complete destruction of primary au-
ditory cortex does not lead to substantial
permanent loss of audiometric sensitivity. It is
more likely, however, for an asymptomatic pa-
tient with old unilateral temporal lobe pathol-
ogy to suddenly become totally deaf with the
onset of the opposite contralateral lesion in
the auditory region.

A neuroanatomic distinction between corti-
cal deafness and auditory cortical disorders
has been tentatively offered by Michel et al.
(1980). Recognizing the hazards of such a dichotomy,
they distinguish between lesions of auditory ko-
nicortex (41-50 of Brodmann) and lesions of
pro- and para-konic cortex (22, 52 of Brod-
mann), respectively. While this distinction
may prove useful, naturally occurring lesions
do not typically obey architectonic boundaries
(Michel et al., 1980).

In their article on cortical deafness, Michel
et al. (1980) considered the possibility that the
two syndromes could be differentiated on the
basis of auditory evoked potentials (AEPs).
Several studies (e.g., Jerger et al., 1969; Michel
et al., 1980) have found either totally absent
cortical AEPs or absent late components of
AEP in patients with cortical auditory disor-
ders. However, AEPs have been found to be
present in other cases (Albert et al., 1972 [pure
word deafness]; Assal and Despland, 1973 [au-
ditory agnosia]), and in at least one case of cor-
tical deafness (Adams et al., 1977), normal late
AEPs were found. While results to date are
conflicting, this remains a promising area of
research. Such variability may be due in part
to differentiating pathologies and recording
methods. Michel et al. (1980) offer methodological
suggestions designed to increase comparability
among patients.

Cortical deafness is most commonly seen in
bilateral cerebrovascular disease in which the
course is commonly biphasic with a transient
deficit (often aphasia and hemiparesis) related
to unilateral damage followed by a second
deficit, associated with evident permanent
defa (Jerger et al., 1969, 1972; Adams et al.,
1977; Earnest et al., 1977; Leitester, 1980).
A biphasic course is also typical of cases of au-
ditory cortical disorder.

In cortical deafness, bilateral destruction of
the auditory radiations or the primary auditory
cortex has been a constant finding (Leitester,
1990). The anatomic basis of auditory cortical
disorder is more variable. Although lesions can
be quite extensive (see Oppenheim and
Newcombe, 1978), the superior temporal gyrus
(i.e., efferent connections of Hesch's gyrus) is
frequently involved. Several recent cases (Mo-
tomura et al., 1986; Kuni et al., 1990; Nish-
okola et al., 1993) suggest that generalized au-
ditory agnosia can result from relatively
circumscribed bilateral subcortical lesions that
impinge upon the auditory radiations.

Pure Word Deafness (Auditory Agnosia for
Speech, Auditory Verbal Agnosia)

Patients with pure word deafness are unable to
comprehend spoken language although they can
read, write, and speak in a relatively nor-
mal manner (Buchman et al., 1966; Ackermann
and Mathias, 1969). By definition, compre-

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The term auditory agnosia refers to the im-
paired capacity to recognize sounds in the pres-
ence of otherwise adequate hearing as mea-
sured by standard audiometry. The term has
been used in a broad sense to refer to impaired
capacity to recognize both speech and non-
speech sounds, and in a more narrow sense to
refer to a selective deficit in the recognition of
nonverbal sounds only. If one uses the broader
definition, then the disorder is further subdi-
vided into auditory sound agnosia, auditory
verbal agnosia, and a mixed group with deficits
in both speech and nonverbal sounds. We pre-
fer the more narrow definition, and will discuss
pure word deafness (a selective impairment in
speech-sound recognition) and auditory agno-
sia (selective impairment in recognizing
nonverbal sounds) separately. The term cortical
deffasness generally has been applied to those
patients whose daily activities and auditory be-
tavior indicate an extreme lack of awareness of
auditory stimuli of any kind, and whose audi-
ometric pure tone thresholds are markedly ab-
normal. Receptive (sensorv) amusia refers to
loss of the ability to appreciate various charac-
teristics of heard music. Phonagnosia refers to
the loss of the ability to recognize familiar
persons by voice.

Cortical Auditory Disorder and Cortical Deafness

In the large majority of cases, impairment of
nonverbal sound recognition is accompanied
by some degree of impairment in the recogni-
tion of speech sounds. The relative severity of
these impairments may reflect premotorial
lateralization of linguistic and nonlinguistic pro-
cesses in the individual patient, and may de-
pend upon which hemisphere is more seri-
ously, or primarily, damaged (Ullrich, 1976).
Terminologic confusion has arisen with regard
to these "mixed" forms, with such terms as
"cortical auditory disorder" (Kanshelpsky et al.,
1973; Miocci, 1982), "auditory agnosia" (Oppen-
hemer and Newcombe, 1978; Rosati et al.,
1982), "auditory agnosia and word deaf-
ness" (Goldstein et al., 1975), and "congenital
aphasia" (Landau and Klieflser, 1957; Landau
et al., 1960) all being used to describe similar
phenomena. We will refer to these mixed forms
as "cortical auditory disorders," and will discuss
them together with cortical deafness. Cortical
auditory disorders frequently evolve from a
state of cortical deafness, and as we shall see,
it is often difficult to define a clear separation
between the two.

Patients with these disorders have difficulty
recognizing auditory stimuli of many kinds,
verbal and nonverbal (Vignolo, 1969; Lher-
mitte et al., 1971). Aphasias, if present, are

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after sustaining severe closed head injury. They
found that, despite efforts to control for these
variables, living things remained selectively im-
paired relative to nonliving things. Their data
do not allow them to specifically discern the lo-
cus of impairment (i.e., whether it is in a cat-
ergically organized semantic system or within
a modularly organized visual system specialized
for the recognition of living things), but they
concluded that some level of visual or semantic
representations specific to living things ap-
ppeared to be involved.

The patient P.S.D. reported by Damasio et al.
(1990) showed a similar, though apparently
more complicated dissociation. This patient
was able to visually recognize man-made tools,
though his recognition of most "natural" stim-
uli was less than 30% accurate. At the same
time, he showed normal recognition of some
natural stimuli (e.g., body parts) and poor
recognition of some man-made stimuli (e.g.,
musical instruments).

Recently, computational models have been
applied to the problem of category specificity in
visual recognition and naming (Farah and
McKearsland, 1991; Small et al., 1995; Devlin
et al., 1998). In these accounts, stimuli are rep-
resented by sets of perceptual features that are
then lesioned in an attempt to produce results
analogous to human neurological syndromes.
Although a detailed review of these models is
beyond the scope of this chapter, the basic re-
sult that emerges from these simulations is that
damage to feature-based computational mod-
els can accurately simulate category-specific
naming and recognition deficits without having
to postulate hierarchical or categorical orga-
nization within the semantic system itself.

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Agnosia

Comprehension often drops suddenly when the topic is changed. Words embedded in sentences are more easily identified than isolated words. Slowing the presentation rate of words in sentences sometimes facilitates comprehension.

Most studies of patients with pure word deafness have emphasized the role of auditory-perceptual processing in the genesis of the disorder (Jüger et al., 1969; Kaszelnikow et al., 1973; Albert and Bear, 1976; Auerbach et al., 1982; Mendez and Geehan, 1988). Temporal resolution (Albert and Bear, 1974), and phonemic discrimination (Chocholle et al., 1975; Denes and Semenza, 1975; Saffran et al., 1976; Nakashiki et al., 2001) have also received attention. In an exceptionally detailed case report and literature review, Auerbach et al. (1985) suggest that the disorder may take two forms: (1) a prephonemic temporal auditory acuity disturbance associated with bilateral temporal lesions, or (2) a disorder of phonemic discrimination attributable to left hemisphere damage.

Albert and Bear (1974) suggested that the problem in pure word deafness is one of temporal resolution of auditory stimuli rather than specific phonetic impairment. Their patient demonstrated abnormal long click-fusion thresholds (time taken to perceive two clicks as one), and improved in auditory comprehension when speech was presented at slower rates. This may lessen the impact of abnormally slow temporal auditory analysis or may allow the patient to reconstruct the message strategically (Neisser, 1967). Saffran and colleagues (1976) showed, however, that informing their patients of the nature of the topic under discussion (indicating the category of words to be presented or giving the patient a multiple-choice array just before presentation of words) significantly facilitated comprehension. Words embedded in a sentence were better recognized, particularly when they occurred in the latter part of the sentence. Whereas a temporal auditory acuity disorder was likely present in Albert and Bear’s (1974) patient, the patient of Saffran et al. (1976) displayed linguistic discrimination deficits that appeared to be independent of a disorder in temporal auditory acuity.

Several studies have reported brainstem and cortical auditory evoked responses in patients with pure word deafness (see Michel et al., 1980, for review). Brainstem evoked potentials are almost universally reported as normal, suggesting normal processing up to the level of the auditory惊. In this review, emphasis was placed on cortical evoked potentials since they are more consistently abnormal in pure word deafness. Most studies report increased latencies and reduced amplitudes of both 100-msec and 200-msec auditory evoked responses to pure words. The dissociation of the evoked potential response to monosyllabic words and monosyllabic pseudowords suggests that there may be a deficit in processing the syllabic structure of the stimulus. This finding is consistent with the hypothesis that pure word deafness is a disorder of phonological processing.
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Disturbed (Buchman et al., 1986). Some patients may recognize foreign languages by their distinctive prosodic characteristics, and others can recognize who is speaking, but not what is said; this suggests preserved ability to comprehend paralinguistic aspects of speech. Coselet and colleagues (1984) described a word-deaf patient who showed a remarkable dissociation between the comprehension of neutral and affectively intoned sentences. It was asked to point to pictures of males and females depicting various emotional expressions. When verbal instructions were given in a neutral voice, he performed poorly. When instructions were given with affective intonations appropriate to the target face, he performed significantly better and at a level commensurate to his performance with written instructions. This patient had bilateral destruction of primary auditory cortex with sparing of auditory association cortex, which suggests at least some direct contribution of the auditory radiations directly to the latter without initial decoding in Heschl's gyrus (Coselet et al., 1984). These authors speculate that one reason why patients with pure word deafness improve their auditory comprehension when lipreading is that face-to-face contact allows them to take advantage of visual cues (gesture and facial expression) that are processed by different brain systems. An alternative explanation is that lipreading provides information about place of articulation, a linguistic feature that is markedly impaired at least in the bilateral cases (Auerbach et al., 1989). In either instance, the finding of preserved comprehension of paralinguistic aspects of speech further reinforces the notion that comprehension of speech and nonspeech sounds may represent dissociable abilities.

There is evidence that unilateral left-sided lesions, particularly those producing Wernicke's aphasia with impaired auditory comprehension, are also associated with impaired ability to match nonverbal sounds with pictures (Vignolo, 1969). However, resulting errors are almost exclusively semantic, not acoustic, and thus do not suggest that unilateral left hemispheric temporal lobe damage produces a perceptual-discriminative sound recognition disturbance. For this reason, the finding of impaired ability to discriminate nonverbal speech sounds in a patient with pure word deafness suggests bilateral disease, even in the absence of other neurological evidence of bilaterality. Since many of these patients have, by history, successive strokes, the primary and secondary side of damage may be important in producing a picture dominated either by pure word deafness or by auditory sound agnosia (Ullrich, 1975).

Although distinctions have been made between basic defects in auditory perception and deficits in linguistic processing, a number of studies of pure word deafness have analyzed the defect in terms of the apperceptive-associative distinction so prominent in discussing visual agnosia (Folster and Rose, 1986). It has been suggested that word deafness may represent the apperceptive counterpart of the very rare and ill-defined disorder called "pure word meaning deafness," in which the patient can hear and repeat words, but does not know their meaning (Corballis, 1994; Franklin et al., 1996).

Auditory Sound Agnosia (Auditory Agnosia for Nonspeech Sounds)

Auditory agnosia for nonspeech sounds is by far more rare than pure word deafness. This may be because such patients are less likely to seek medical advice than those with a disorder of speech comprehension, and also because nonspecific auditory complaints may be discounted when pure tone audiometric and speech discrimination thresholds are normal. This is unfortunate, since normal or near-normal audiometric evaluation does not rule out the possible role played by primary auditory perceptual defects (Goldstein, 1974; Buchtel and Stewart, 1969).

Vignolo (1969) argued that there may be two forms of auditory sound agnosia: (1) a perceptual-discriminative type associated mainly with right hemisphere lesions involving Brodmann's area 41, 42, and 35, and (2) an associative-semantic type associated with lesions of the left hemisphere involving Brodmann's area 37 and 20, and closely linked to Wernicke's aphasia. This anatomic distinction is by no means settled. The former group makes predominantly acoustic (e.g., "mean whistling" for birdsong) errors on picture-

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sound matching tasks, while the latter group makes predominantly semantic (e.g., "train" for automobile engine) errors. This division follows the original classification of Kleet (1928), who distinguished between the ability to perceive isolated sounds or noises and the inability to understand the meaning of sounds. It also resembles the apperceptive-associative dichotomy made by Lissauer (1890). In the verbal sphere, the analogous distinction is between pure word deafness (perceptual-discriminative) and transcortical sensory aphasia (semantic-associative).

Few stable cases of "pure" auditory sound agnosia have been reported in the literature (Nielsen and Sulk, 1939; Worts and Pfiffer, 1948; Spreen et al., 1965; Albert et al., 1972; Fujii et al., 1980; Schneider et al., 1994). Sometimes a patient evokes into auditory sound agnosia from a more generalized agnosia for both verbal and nonverbal sounds (Motomura et al., 1988), and occasionally a patient evokes from an auditory sound agnosia into an auditory recognition defect that encompasses speech sounds and other auditory stimuli (Kaga, 1999).

The patient of Spreen et al. was a 65-year-old right-handed male whose major complaint was that he has been a "half-hemiparetic episode of "nerves" and headaches. Audiometric testing demonstrated moderate bilateral high-frequency loss and speech reception thresholds of 12 dB for both ears. There was no aphasia. The outstanding abnormality was the inability to recognize common sounds; understanding of language was fully retained and there were no other auditory defects. Sound localization was normal. Scores on the pitch subtest of the Seashore Tests of Musical Talent were at chance level. The patient claimed no experience or talent with music and refused to cooperate with further testing of musical ability. The patient could match previously heard but misidentified sounds with one of four tape-recorded choices, suggesting an associative defect. Postmortem examination revealed a sharply demarcated old infarct of the right hemisphere involving the superior temporal and angular gyrus, as well as a large portion of the inferior temporal, inferior parietal, and middle frontal, and long and short gyri of the insula. This case represents one of the few examples of auditory sound agnosia with unilateral pathology. The lesion is too large to allow for precise anatonioclinical correlation. Other cases with unilateral pathology include those reported by Fujii et al. (small posterior right temporal hemorrhagic lesion that involved the middle and inferior temporal gyri), Nielsen and Sulk (right thalamus and parietal lobe), and Worts and Pfiffer (large lesion of the right temporoparietal-occipital junction). The case reported by Fujii et al. (1990) is informative because he was completely free of aphasic symptoms and his lesion was gratifyingly small. In this patient, pure tone audiometry was within normal limits in spite of a 30 dB high-frequency hearing loss in the left ear. He showed marked suppression of the left ear during dichotic listening tests involving digits and words. Brainstem auditory evoked responses (BAERs) were normal and sound localization was intact. The patient was selectively impaired in identification of nonspeech sounds, and his errors consisted primarily of acoustic confusions ("sound of railroad crossing" for telephone ring). His agnosia cleared by the 16th post-stroke day.

Albert et al. (1973) described a patient with auditory sound agnosia with minimal dysphasia. Clinical evidence suggested bilateral involvement. The patient was able to attach meaning to words-sounds, but not to nonverbal sounds. Albert et al. also demonstrated the marked extinction of the left ear to dichotic listening, impaired perception of pitch, loudness, rhythm, and time; and abnormally delayed and attenuated cortical AEs, worse on the right. They concluded that the sound agnosia in their patient resulted from an "ability to establish the correspondence between the perceived sound and its sensory or motor associations" (associative defect), and suggested that the dissociation between verbal and nonverbal sound recognition in their patient reflected different processing mechanisms for linguistic and non-linguistic aspects of acoustic input.

Sensory (Receptive) Amusia

Musical ability is a complex domain of functions in which selective impairments can occur after brain damage. Several distinct disorders
have been identified, including vocal amusia, loss of skilled instrumental ability (instrumental amusia, McFarland and Fortin, 1982), loss of the ability to read write music (musical alexia and agraphia; Brust, 1980; Midorikawa and Kawamura, 2000), impaired recognition of musical (receptive amusia; Procopis, 1983; Piccirilli et al., 2000; Schuppert et al., 2000), and disorders of rhythm (Berman, 1981). Analysis of the patient with reported musical impairments should take into account the multicomponential nature of musical abilities.

The subject of amusia has been reviewed in detail by Wertheim (1969), Critchley and Henson (1977), and Gates and Bradshaw (1977). Sensory (receptive) amusia refers to an inability to appreciate various characteristics of heard music. It occurs to some extent in all cases of auditory sound agnosia, and in most cases of aphasia and pure word deafness, but can occur independently of other deficits (Piccirilli et al., 2000). As is the case with auditory sound agnosia, the loss of musical perceptual ability may be underreported because a specific musical disorder rarely interferences with everyday life. A major obstacle to systematic study of acquired amusia is the extreme variability of pre-illness musical abilities, interests, and skills. It was Wertheim's (1969) opinion that receptive amusia corresponds more frequently with a lesion of the left hemisphere, while expressive musical disabilities are more apt to be associated with right hemisphere dysfunction. Recent evidence indicates that cerebral organization of musical ability is dependent on degree of experience, skill, and musical sophistication. Musically skilled and trained individuals may be more likely to perceive music analytically and to rely more heavily on the dominant hemisphere. Dichotic listening studies show that the right hemisphere plays a more important role than the left in the processing of musical and auditory sound patterns (Blumstein and Cooper, 1974; Gordon, 1974). However, the left hemisphere appears to be of major importance in the processing of sequential (temporally organized) material of any kind, including musical series. According to Gordon (1974), melody recognition becomes less of a right-hemisphere task as time and rhythm factors become more important for distinguishing tone patterns (see also Lavrov, 1980), although it has been argued that rhythm and interval processing may be vulnerable to right hemisphere damage (Schuppert et al., 2000). Such conclusions contribute to a lack of definition of the entity of receptive amusia and the difficulty of localizing the deficit to a particular brain region. Further complicating the picture is the fact that pitch, harmony, timbre, intensity, and rhythm may be affected to different degrees in combinations in the individual patient. Furthermore, there is evidence that aspects of musical denotation (the so-called real-world events referred to by lyrics) and musical connotation (the formal expressive patterns indicated by pitch, timbre, and intensity) are selectively vulnerable to focal brain lesions (Gardner et al., 1977). For instance, on tests of musical denotation, right hemisphere–damaged patients perform well on items where acquaintance with lyrics is required; in contrast, aphasics with anterior lesions perform better than both right hemisphere–damaged patients and aphasics with posterior lesions (items where knowledge of lyrics is unnecessary). (Incidentally, Benton [1980] reports that aphasics with posterior lesions and comprehension disturbance are also more impaired among aphasics on tests of face recognition, another ostensibly "configural" task. On tests of musical connotation, right hemisphere–damaged patients do better in matching sound patterns to temporally sequenced designs than to simultaneous gestaltes. Aphasics with posterior lesions perform relatively well on tests of musical connotation. Comprehensive tests of musical ability that separate these many subskills are now available (Schuppert et al., 2000).

Perezt and colleagues (1994) applied comprehensive nonverbal auditory testing to two patients with bilateral lesions of auditory cortex. In their patients, the perception of speech and environmental sounds was spared, but the perception of tones, prosody, and voice was impaired. On the basis of these behavioral dissociations, they argue that music processing is distinct from processing of speech or environmental sounds (Piccirilli et al., 2000). Their data led them to argue for a task- and process-specific approach to the analysis of cases of auditory agnosia. They suggest that nominally auditory tasks be broken down into their functional subcomponents and that more extensive component-based analysis of auditory processing deficits is warranted. For example, they distinguish between processes involved in the recognition of specific voices or musical instruments (which are timbre-dependent), and processes involved in recognition of tunes (which are pitch-dependent). The notion that nominally distinct classes of auditory material (e.g., melodies, prosody, and voice) share common processes may be critically important in developing a functional taxonomy of auditory recognition disorders in general, and of amusia in particular (Schuppert et al., 2000).

This suggestion points out certain significant deficiencies in the evaluation of amusia patients. Although theories linking brain function to music perception have long been available (Hécaen, 1962; Bever and Chiarello, 1974), such theories do not often contain sufficient process specificity to guide the clinical evaluation of amusia patients. Thus, for example, relatively little is known about the musical features that will be most informative in constructing a neuropsychological model of music perception. Another obstacle to systematic study of acquired amusia is the variability of pre-illness musical abilities, interests, and experience (see Wertheim [1969] for a system of classifying musical ability level). The cerebral organization of musical perception has been suggested to be dependent upon one of these pre-illness characteristics (Bever and Chiarello, 1974).

Paralinguistic Agnosia: Auditory Affective Agnosia and Phonagnosis

Heilman and colleagues (1975) showed that patients with right temporoparietal lesions and the neglected syndrome were impaired in the comprehension of affectively intoned speech, but showed normal comprehension of speech content. Patients with left temporoparietal lesions and fluent aphasia comprehended both affective (paralinguistic) and content (linguistic) aspects of speech normally. Whether this speech deficit represents a true agnosia remains to be determined, since auditory sensory-perceptual skills were not assessed. It is possible that auditory affective agnosia is a variant of auditory sound agnosia, i.e., that it represents a category-specific auditory agnosia.

Studies by Van Lancker and associates (Van Lancker and Kreiman, 1988; Van Lancker et al., 1985, 1989) have revealed another type of paralinguistic deficit after right hemisphere disease. In their studies, patients with unilateral right hemisphere disease showed deficits in discriminating and recognizing familiar voices, while patients with left hemisphere disease were impaired only on a task that required discrimination between two famous (but not personally familiar) voices. The CT evidence suggested that right parietal damage was significantly correlated with voice recognition impairment, while temporal lobe damage in either hemisphere led to deficits in voice discrimination. The authors refer to this deficit as "phonagnosia," but, like auditory affective agnosia, it remains to be seen whether it is truly agnosic in nature. A recent functional imaging study revealed that a distributed cortical network may be involved in the processing of familiar visual and vocal stimuli (Shah et al., 2001). In this MR study, subjects viewed familiar and unfamiliar faces and listened to personally familiar and unfamiliar voices. Changes in neural activity associated with stimulus modality irrespective of familiarity were observed in regions known to be important for face recognition (fusiform gyrus bilaterally) and voice recognition (superior temporal gyri bilaterally), while familiarity of voices and voices (relative to unfamiliar voices) was associated with increased neural activity in the posterior cingulate cortex, including the retrosplenial cortex. Although the status of these defects vis-à-vis the concept of agnosia is uncertain, the discovery of seemingly specific impairments in the comprehension of affectively intoned speech and speaker identity is important, as paralinguistic abilities may be spared in cases of pure word deafness (Coslet et al., 1984). As indicated above, patients with pure word deafness frequently report that they are able to recognize the speaker of the message and, less frequently, the language in which it is transmitted. These findings lend further support to the idea that linguistic and paralinguistic
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discriminative defects, while Delay and others use the term to denote defects of object identification. It is clear that defects in two-point discrimination, point localization, and position sense can impair tactile form perception, and thus object identification without producing concomitant defects in sensitivity to light touch, temperature, or pain (Gans, 1916; Campera, 1925; Corkin, 1978). However, significant defects in discriminative ability need not accompany disorders of tactile identification (Corkin, 1978). Thus, clinical data suggest that tactile discrimination and identification are dissociable. Unfortunately, the vast majority of the physiological and anatomic data on somatosensory agnosia have come from animal research using almost exclusively discrimination, rather than identification, paradigms.

With these considerations in mind, we will use the term cortical tactile disorders to refer to a diverse spectrum of defects in somatosensory discrimination or recognition of distinct object quaestions. (1) Impaired two-point tactile agnosia for those rare cases in which there is an inability to identify the nature of tactually presented objects despite adequate sensory, attentional, intellectual, and linguistic capacities. Although debatable, we will discuss astereognosis as an apperceptive form of tactile agnosia, recognizing that it represents a failure of complex perceptual processing that has, as an outcome, an impairment in tactile object recognition ability.

Before discussing disorders of tactile recognition, some comments about the functional anatomy of the somatosensory systems are necessary. An exhaustive review of this vast literature will not be undertaken here; the interested reader is referred to excellent reviews by Hécaen and Albert (1978), Corkin (1978), Mountcastle and Powell (1959a, 1959b), and Werner and Whisler (1973).

Two relatively distinct somatosensory systems have been identified. One is the spinothalamic system: cutaneous nerve endings → spinothalamic tract → reticular formation → thalamic nuclei → superior bank of Sylvian fissure (Hécaen and Albert, 1978; Brodal, 1981). In addition, SI projects heavily to area 5 (superior parietal lobule) (Jones and Powell, 1969a, 1969b; Corkin, 1978), which is important for motor pursuit of motivationally relevant targets in extrapersonal space (Mountcastle et al., 1975).

Thus, the functional interconnections of cortical somatosensory areas involve regions that, from numerous other studies, have been found to subserve motor, proprioceptive, and spatial functions. The existence of such a complex system in the human brain is important for intentional, spatially guided motor movements that bring us into contact with tactile stimuli. Reciprocal connections between somatosensory, motor, proprioceptive, and spatial components of the system provide the mechanisms through which regulation of the perceptual act can be achieved. The complex functional organization of the somatosensory systems underscores the idea that perception is an active process and involves more than the mere passive processing of environmental input.

Although patients with lesions of the afferent somatosensory pathways frequently cannot identify tactually presented objects, this is due to a severe sensory loss, sometimes referred to as "stereognosia." Lesions of the primary visual and auditory areas produce specific disorders of sensation that can vary in severity depending on the extent and location of the lesion (Rush, 1965, and Semmes, 1974) selectively affecting visual and auditory areas results in cortical blindness or deafness, respectively. In contrast, disorders of sensation for touch, temperature, pain, and vibration are rare following cortical lesions (Hécaen and Albert, 1978). Redundancy in representation seems to be an especially important characteristic of somatosensory systems. Paul et al. (1972) explored units in anatomic subdivisions of SI, and found multiple representations of the monkey’s hand, one in each subdivi- sion (see also Mountcastle and Powell, 1959a, 1959b; Powell and Mountcastle, 1959).

Randolph and Semmes (1974) selectively ablated each of the SI subregions (3b, 2, 1). Area 3b excisions resulted in impairment of all aspects of tactile discrimination learning. Lesions of area 1 produced loss of hair and touch discrimination, spared complex-concave and square-diamond shape discrimination. The opposite pattern was seen with area 2 lesions. Thus, the hand appears to be represented and re-represented within specific subdivisions of somatosensory cortex according to sensory “submodality.”

The notion of sensory modularity dates back at least to von Frey (1895), who divided the tactile sense into light touch, pressure, temperature, and pain sensitivity. Head (1918) di-
vided sensory functions into three categories: (1) recognition of spatial relations (passive movement, two-point discrimination, and point localization), (2) relative sensitivity to touch, temperature, and pain, and (3) recognition of similarity and difference (size, shape, weight, and texture). Submodalities may be selectively impaired, while others are spared, by circumscribed cortical lesions. Head's (1918) framework, for example, suggests that discriminatory defects are accompanied by defects in the discrimination of texture and weight, but not by impaired perception of spatial relations, touch, temperature, or pressure (see Corkin, 1978). On the basis of studies of recovery from peripheral nerve injuries, Head and colleagues (1905) distinguished between "protopathic" and "epipptic" sensation. The epipptic system serves focal point sensibility, while the protopathic system is more diffuse. The protopathic-epipptic distinction has been widely accepted by anatomists and physiologists (Rose and Woolsey, 1949; Mountcastle, 1961), but unlike Head, these authors have emphasized the anatomic implications of this distinction at the cortical and thalamic levels, rather than at the peripheral level (Hécaen and Albert, 1978, p. 279 ff). As implied previously, the epipptic aspects of touch are more directly subserved by the medial lemniscal-S1 system, while the protopathic dimension relates more closely to the functions of the bilaterally represented S1, although there is considerable functional overlap between the two systems.

Cortical Tactile Disorders

The brief review of the somatosensory systems has been designed to emphasize the complexity of this sensory modality and to enable the reader to anticipate the enormous variability in presentation among patients suffering from tactile recognition and identification disorders. Historically, there have been two views regarding the nature and functional localization of disorders of tactile sensation. The first, more traditional, view is that sensory defects are associated with the contralateral primary somatosensory projection area in the postcentral gyrus (Head, 1920). The other perspective is that more diffuse aspects of cortex (e.g., posterior parietal lobe) are involved in somatosensory perception (Semmes et al., 1960).

In a series of studies, Corkin and colleagues (Corkin, 1964; Corkin et al., 1970, 1973) administered quantitative tests of pressure sensitivity, two-point discrimination, point localization, position sense, and tactile object recognition to patients who had been operated on for relief of focal epileptic seizures. Lesions in the contralateral post-central gyrus produced the most severe disorders of cortical tactile sensation. Also, clear demonstration was made of the existence of bilateral sensory defects associated with a unilateral cortical lesion, as had been previously reported by Semmes et al. (1960) and Oppenheim (1906).

Corkin found that the most severe defects occurred in patients whose lesions encroached on the hand area. This is consistent with the findings of Roland (1976) in his studies of tactile shape and size discrimination impairment with focal cortical lesions. Corkin et al. (1970) also found that disorders of tactile object recognition were restricted to the contralateral hand in patients with lesions that involved the hand area in S1. Importantly, defects of tactile object recognition were always associated with significant defects in pressure sensitivity, two-point discrimination, and other elements of sensory function. Patients with parietal lobe lesions sparing SI did not show object identification disturbances.

Twenty of 50 patients with parietal lobe involvement showed additional sensory defects ipsilateral to the damaged hemisphere (Corkin et al., 1970). This effect was found with equal frequency after acute and chronic hemisphere excisions, in contrast to previous studies that had found the incidence of ipsilateral sensory impairment to be much more frequent following left hemisphere damage (Semmes et al., 1960). Differences in the extent of lesions in the samples used by Corkin et al. (circumscribed cortical lesions) and Semmes et al. (penetrating missile wounds) may account for some of these discrepancies. An important anatomic fact is that, in patients with bilateral sensory defects of the hand, the postcentral hand area needs not be involved (Corkin et al., 1973). The area of damage implicated in these patients was tentatively offered as SII. In summarizing these data, Corkin (1975) suggested that unilateral SII hand area lesions produce severe contralateral sensory deficits, while unilateral SII lesions may produce milder defects that affect both hands.

There is growing evidence of hemispheric specialization for certain higher somesthetic functions. Data on this issue can be found in cerebral laterality studies, examinations of patients following brain biopsy, and in studies of performance on complex somatosensory tasks after unilateral hemispheric lesions (Milner and Taylor, 1972; Corkin, 1978; Hécaen and Albert, 1978). While laterality studies have failed to show hemispheric specialization for elementary somesthetic functions such as pressure sensitivity (Fennell et al., 1967), vibration sensitivity (Seiler and Rickert, 1971; cited in Corkin, 1978), two-point discrimination (McCall and Cunningham, 1971), or point localization (Semmes et al., 1960; Weinstein, 1966), results of complex sensory tasks requiring spatial exploration of figures or fine temporal analysis reveal evidence of hemispheric specialization. The left hand-right hemisphere combination appears especially proficient at tasks in which a spatial factor is important, such as in exploring braille (Rudel et al., 1974) or perceiving the spatial orientation of tactually presented rods (Benton et al., 1973). Results from studies of split-brain patients (reviewed in detail by Corkin, 1978) are consistent with these conclusions: the left hand-right hemisphere combination is better able to perform complex, spatially patterned discriminations, although the right hand hemisphere can succeed if familiar stimuli are presented, if a small array of objects is involved, or in other situations in which linguistic processing can be effectively used (Milner and Taylor, 1972).

Thus, patients with right hemisphere disease do worse than left hemisphere-damaged patients on tasks requiring the perception of complexly organized spatial stimuli, although any patient with elementary somatosensory dysfunction, regardless of hand, can be expected to do poorly with that hand (Corkin, 1978). Semmes (1965) has identified a group of patients with profound sensory tactile impairment who fail tests of shape-discrimination. These patients were unimpaired in roughness, texture, and size discrimination, but showed profound impairments on tests of spatial orientation and route finding. These patients suffered from lesions of the superior parietal lobe. Semmes concluded that there is a "non-tactual" factor in these discriminative defects that transcended sensory modality. According to her view, what is spatial for vision is represented in touch by the temporal exploration of object qualities. Teuber (1955a, 1955b) interpreted the difficulty as a special form of spatial disorientation, rather than one of "agnosia for shape." To summarize, no hemispheric specialization appears to exist for elementary somatosensory function, although there is growing evidence that the right hemisphere is more strongly involved in processing the highly spatial character of some tactile discrimination and identification tasks. Postcentral gyrus lesions frequently result in severe and long-lasting defects in using the contralateral hand, while lesions of SII result in less severe, bilateral defects. A general conclusion from this extensive and complex literature is that the central regions (so-called sensorimotor cortex) are more directly involved in elementary somatosensory function, while complex somatosensory tasks involving strong spatial or motor exploratory components involve additional structures posterior to the sensory motor region (Corkin, 1978). This distinction makes it possible to see a higher somatosensory disorder in the absence of elementary sensory loss. Whether this higher disorder deserves to be called "agnosia" is a subject to which we now turn.

Tactile Agnosia

The patient with tactile agnosia cannot appreciate the nature or significance of objects placed in the hand despite elementary somatosensory function, intellectual ability, attentional capacity, and linguistic skill adequate to the task of object identification. The terms astereognosis and pure astereognosis have been sometimes used synonymously with tactile agnosia, and sometimes used to describe basic defects in the appreciation of size, shape, and texture. Delay (1935) asserted that astereognosia...
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agnosia was a complex disorder comprised of amorphognosia, agraphognosia, and tactile agnosia. In our view, agnosia, as it has been described in the literature, essentially refers to "apraxic agnosia," that is, we will use the term tactile agnosia for cases in which a deficit of tactile object recognition exists without corresponding sensory-perceptual defects.

Clinical case reports of pure agnosia are rare (Raymond and Eggie, 1963; Buxton, 1915; Campana, 1925; Hecaen and Dam, 1945; Newcombe and Ractcliffe, 1974). Frequently, obvious sensory defects do appear at some point in the clinical course of these patients, though not necessarily coincident with the identification disturbance. The agnostic patient frequently has defects limited to one hand, usually the left, although patients with defects limited to the right hand have been reported (Hecaen and Dam, 1945).

In some cases, the symbolic hand can eventually achieve recognition of the object, but only after protracted linguistic analysis of the separate features.

Many agnostic patients do not normally palpate the object when it is placed in the hand for identification (Oppenheim, 1906, 1911). This suggests a defect in the mechanism through which tactile impressions are collected to form an integrated percept of the whole object, or a defect in stored structural tactile representations. Motor and sensory information is highly integrated in the act of palpating an object; motor commands are issued that direct the hand in ongoing exploration. In a series of experiments using regional cerebral blood flow (rCBF) during tactile agnostic testing in humans (Roland and Larsen, 1976), have shown that local rCBF increases occur most strongly in the contralateral sensorimotor hand area and the premotor region. Although sensorimotor integration and proprioception are crucial components in tactile identification, it should be noted that the motor component probably has a complex role and is not obligatory in any simple sense. This conclusion is warranted by two clinical facts: (1) motor paralysis does not necessarily cause tactile identification disturbances (Caselli, 1991b), and (2) objects can often be identified if they are passively moved across the subject’s hand, independent of active manipulation. Still, the fact that true agnoses do not palpate objects suggests that elementary sensory function is not actively brought to bear, nor is it adequately integrated with motor information, in the perceptual processing of the stimulus.

Evidence for an associative defect exists when elementary somatosensory defects are either absent or too mild to account for a tactile object recognition disturbance and when the patient can draw or match tactually presented stimuli. For example, the patient of Hecaen and Dam (1945) who could not name an object placed in the hand could draw an accurate picture of the object and could then name the picture. The patients of Newcombe and Ractcliffe (1974) could tactually match to sample, even though they had a disturbance in recognition of the nature of objects.

In other forms of agnosia, there has been significant debate regarding the existence of true tactile agnosia. Three general explanations have been proposed. The argument that there is a familiar hand disturbance of tactile object identification can be traced to defects of elementary somatosensory dysfunction. The second states that the defect is not an agnosia, but instead represents a modality-specific anoma. Third, there are those who do not deny the existence of higher defects of tactile identification in the context of normal elementary somatosensory function, but say that the defect of function in agnosia is spatial and supramodal, involving both tactile and visual disturbances. Because one of the hallmarks of the agnosia concept is its modality specificity, this third viewpoint destroys the notion that tactile object identification disturbances are agnosic in nature. Each of these views is capable of handling some, but not all, of the data. We will briefly examine the status of each of these arguments below.

The potential role of subcortical somatosensory defects in producing disorders of tactile identification has been raised by several authors (Head and Holmes, 1911; Bay, 1944; Corina et al., 1970). Bay (1944) stated that most putative cases of tactile agnosia had been inadequately tested for elementary somatosensory dysfunction, and speculate that thresholds and deficits in performing complex sensory discriminations. Head and Holmes (1911) also stressed the importance of inconstant thresholds, and found that rapid local fatigue and abnormal persistence of sensations frequently brought to bear, nor is it adequately integrated with motor information, in the perceptual processing of the stimulus.

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As in other forms of agnosia, there has been significant debate regarding the existence of true tactile agnosia. Three general explanations have been proposed. The argument that there is a familiar hand disturbance of tactile object identification can be traced to defects of elementary somatosensory dysfunction. The second states that the defect is not an agnosia, but instead represents a modality-specific anoma. Third, there are those who do not deny the existence of higher defects of tactile identification in the context of normal elementary somatosensory function, but say that the defect of function in agnosia is spatial and supramodal, involving both tactile and visual disturbances. Because one of the hallmarks of the agnosia concept is its modality specificity, this third viewpoint destroys the notion that tactile object identification disturbances are agnosic in nature. Each of these views is capable of handling some, but not all, of the data. We will briefly examine the status of each of these arguments below.

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patient could demonstrate the use of tactually presented objects. Naming errors were frequently semantic confusions. The authors interpreted the deficit as a "bilateral tactile aphasia," and suggested that it represents the tactile analogue of "optic aphasia" (Lhermitte and Beauvois, 1973). The third difference is that Geschwind and Kaplan's patient showed normal tactile exploration of objects, while patients with aphasic tactile agnosia (agno-
asia) show deficient palpation of objects, characterized either by a reluctance to manipulate the object or by a stereotypic pattern of manipulation that is independent of specific object qualities.

Supramodal Spatial Defects

Some patients with tactile recognition disorders also have profound defects in spatial localization, route-finding, and other visuospatial tasks (Semmes, 1965; Corbin, 1978). In concluding her review of somatosensory function, Corbin (1978) stated that it is "possible to observe an impairment of high tactile functions in an individual whose elementary sensory status is preserved. It is inappropriate, however, to call this impairment an agnosia, because the higher-order deficits seen are not specific to somesthesia" (p. 145). Recent evidence suggests that tactile recognition may be dependent on spatial and visual processing, perhaps representing the assembly of an object's structural description (Platz, 1996; Saetti, 1999; Zangaladze et al., 1999). This is a persuasive and important argument, but should not be taken to mean that tactile agnosia as a modality-specific entity does not exist in some patients. It is possible, for example, to suffer from an isolated deficit in tactile shape perception without the contribution of apparent spatial disability (Redd et al., 1998). It is likely that, in many cases, large lesions involving parietal cortex and underlying white matter affect neural systems involved in supramodal spatial ability in addition to specifically involving the second somatosensory system. What may result from this kind of lesion is a sort of mixed deficit in which somatosensory and spatial factors combine in unspecified amounts. The fact that such a complex disorder exists does not negate the possibility that, with more restricted lesions, a parietal, modality-specific defect corresponding to tactile agnosia will result.

Although the anatomically and clinical evidence is far from clear, it seems reasonable to distinguish four defects of somatosensory recognition: cortical tactile disorders, agnosia, and disorders of tactile naming secondary to tactile-verbal disconnection. Cortical tactile disorders involve defects in basic and intermediate somatosensory function, the end result of which will be pervasive somatosensory impairment in addition to defects in TOT. We believe that agnosia is a more specific defect, that it deserves a designation as an apperceptive tactile agnosia (but see Tranel, 1991), and that it is primarily caused by a lesion in the functional system preserved by the middle third of the postcentral gyrus (the hand area) and its cortical and subcortical connections. Tactile agnosia, in contrast, seems to result from parieto-temporal lesions that primarily involve S1 insuperradiation (Correll, 1991). Observations of left hemisphere damage in patients with tactile agnosia have led to the hypothesis that a lesion in S1 or between S1 and S2 is necessary to account for anterior somatosensory deficits (Endo et al., 1992).

Understanding complex somatosensory function in the individual patient requires a systematic neurophysiological evaluation of the task of perceptual object identification as well as an evaluation of elementary and intermediate somatosensory function (Caselli, 1991a). When an object is palpated, sensory and proprioceptive cues received by postcentral gyrus interact with the premotor region to direct a series of coordinated movements necessary to construct a tactile image of the object. Most TOT tasks contain components that could be described as sensory, spatial, proprioceptive, constructive, and motor. The functional interconnections between S1, premotor region, and more posterior portions of the parietal cortex highlight the challenges in functionally separating these task dimensions in tasks of TOT.

AGNOSIA AND CONSCIOUS AWARENESS

Despite a profound disability in direct identification of objects or faces, many agnosics are able to demonstrate some knowledge about the stimulus if appropriate tests of recognition are used. Generally speaking, such tests have the common characteristic of not requiring the patient to make direct conscious reference to stimulus identity. Instead, the task is structured such that knowledge is demonstrated in an indirect or implicit way.

Dissociations of this type have received substantial discussion in the memory and attentional literatures (see Chapters 13, 18, and 19). For example, it has been shown that severely amnesic patients are able to acquire new motor or perceptual skills and can demonstrate intact perceptual and conceptual priming of previously presented information (Roediger et al., 1994; Meade and Keane, 1997). Similarly, patients with hemispatial neglect are capable of perceiving unattended information and, in some cases, engaging in high-level semantic processing of neglected stimuli despite being unaware of their perceptions (McClurey Bernto, et al., 1993; Farah and Feinberg, 1997). Other evidence suggests that patients are capable of lexical access without awareness in acquired alexia (Landis et al., 1980; Saffran, 1986; Caselli and Safran, 1989a), preserved visual identification capacity in hemianopic fields ("blind sight"; Weiskrantz et al., 1974; Weiskrantz, 1980), and preserved semantic priming in Wernicke's aphasia (Milberg and Blumstein, 1981; Blumstein et al., 1982). In all of these examples, evidence exists that the cognitive system is able to process in these patients in a way that is independent of conscious awareness.

The best evidence for covert recognition in agnosia comes from studies of patients with prosopagnosia. In a study of eight patients with prosopagnosia, Byrner et al. (1983) provided the first behavioral evidence of covert recognition when they showed that their prosopagnosic was more easily able to match unrecognizable faces with their real names than with arbitrary names. De Haan et al. (1987a) found that their prosopagnosic, like normal controls, performed same-different judgments more rapidly when the task involved famous faces than when unknown faces were presented. In these two examples, performance benefited from familiarity, even though the patients never recognized specific faces as familiar.

In an elegant series of studies, De Haan et al. (1987a, 1987b) used the face-name interference (FNI) task to explore preserved semantic processing in prosopagnosia. In FNI, subjects are asked to make a semantic classification judgment ("Is this an actor or politician?") when presented printed names. Presented along with the name, but irrelevant for the name classification task, is a face that is (a) the same person as the printed name, (b) from the same semantic category as the printed name, or (c) from a different category. In normal subjects, name classification is slowed by the presence of a face from a different category, presumably based on a stroop-like phenomenon. Prosopagnosics also show this effect, even though they fail to recognize a single face (De Haan et al., 1987a, 1987b). This suggests that, at some level, they can extract semantic information from faces and can thus become distracted by it when they perform the name classification task.
faces, but concentrated on internal features when viewing famous faces. Thus, the manner of visual exploration reveals that, at some level, facial familiarity is detected by the visual system, though it is not reflected in the patient's verbal report.

Similar techniques have been applied to the question of whether prosopagnosics can learn new faces. Sergent and Poncet (1990) asked their prosopagnosic to inspect a series of famous novel faces in preparation for a subsequent recognition task. Afterward, some of these faces (old) were combined with series of new faces, and subjects were asked to perform an old-new discrimination. Although the patient was generally unable to discriminate between old and new faces, face familiarity led to increased accuracy in episodic recognition. Remarkably, the patient was able to directly recognize faces on some trials.

Greve and Bauer (1990) used a variant of Zajonc’s “mere exposure” paradigm to demonstrate covert learning of faces in their patient. The patient was shown a series of faces that were each later paired with a novel distractor for forced-choice recognition and preference judgments. In forced-choice recognition, the patient was asked to indicate which of the two faces had been previously presented. In the preference-judgment task, the patient was asked which of the two faces he liked better. He performed at chance in forced-choice recognition, but liked significantly more of the target faces than predicted by chance. Thus, both of these studies suggest that prosopagnosics can learn some aspects of new faces, provided that familiarity is assessed indirectly.

In a few patients, psychophysiological (Bauer, 1986) and behavioral (Newcombe et al., 1989) measures have failed to reveal evidence of covert recognition (see Buer, 1991 for review). These patients had significant apperceptive defects, which suggest impairment prior to the level of the face-recognition unit.

Thus covert recognition may be a characteristic of associative prosopagnosia.

These findings, and those in the literature on amnesia, alexia, aphasis, neglect, and blindsight, suggest that a substantial amount of perceptual and semantic processing must be intact in these patients prior to or independent of the process that generates contextual or autobiographical recognition (Damasio, 1989; Schacter, 1989). Second, they imply that autobiographical resolution of the nature of a stimulus involves mechanisms that are different from or additional to the mechanisms that process stimulus attributes.

One important issue concerns the implication that such findings have for the functional architecture of object recognition. One of the dominant viewpoints is that such findings implicate the dissociation between stimulus processing and consciousness. Schacter (1989) offers a general account of such dissociations by proposing that implicit and explicit domains reflect parallel, nonoverlapping outputs of early, “modular” cortices dedicated to processing specific types of information. In this view, one set of outputs links the outputs from sensory cortex with motor, verbal, and viscerovisceral response systems that are themselves capable of reaching out cortical activity without conscious or even mnemonic control. Another independent set of outputs links modular cortices with a conscious awareness system that is responsible, in the case of object recognition, for explicit stimulus identification. According to this model, covert recognition in agnosia would result when functional interaction between modular cortices is selectively interrupted or disconnected while modular outputs to motor, verbal, and autonomic response systems remain intact.

This impairment of functional interaction between modular cortices and the awareness system would lead to the kind of deficit one sees in agnosia: a domain-specific impairment in conscious identification without a global impairment in conscious awareness.

An alternative model, not requiring a disconnection mechanism, postulates that dissociations between implicit and explicit forms of object/face identification are the natural result of a damaged object recognition processor (Farah et al., 1993). This general approach was outlined above in the section on prosopagnosia, and is useful because (1) it begins to specify the nature of information processing taking place at different levels of the nervous system, and (2) it supplements lesion work in a way that informs and constrains the analysis of individual cases.

EXAMINATION OF THE PATIENT WITH AGNOSIA

Two basic principles should guide the examination of the agnostic patient. First, care should be taken to rule out the possibility that the recognition disorder is attributable to sensory-perceptual dysfunction, inattention, aphasia, generalized memory loss, or dementia. Second, an extensive analysis of the scope and limits of the patient's deficit is required to characterize its functional locus.

RULING OUT ALTERNATIVE EXPLANATIONS

At the outset, it is important to remember that agnostic recognition failures are basically modality-specific. Patients who exhibit multimodal defects are more likely to be suffering from amnestic syndrome, language disturbance, dementia, or, in rarer cases, generalized impairments in semantic access. The nonaphasic agnostic patient will not usually manifest word-finding difficulty in spontaneous speech, and will generally succeed in generating lists of words in specific categories, in completing open-ended sentences, and in supplying words that correspond with definitions. Except in the rare case of optic aphasia, the agnostic will not be able to identify the mismapped objects by means of circumlocution, or by indicating function. It is thus important to determine whether the patient is able to demonstrate the use of objects not in his or her presence and to follow commands not requiring objects (e.g., salute, wave goodbye, make a fist, etc.) and to require that the patient consciously identify the objects. Thus, intact auditory comprehension indicate apraxia; subsequent failure to demonstrate the use of objects presented on visual confrontation may therefore be apraxic, not agnostic.

In pointing and naming tasks, it is important to be certain that the patient is visually fixating on the objects to be identified and that pointing errors are not due to mislocation in space. Recognition should be examined both in the context of normal surroundings and in the formal test setting, taking care to ensure that the patient is familiar with target objects.

As a start, comprehensive neuropsychological assessment of intellectual skill, memory function, language, constructional/perceptual ability, attention, problem-solving, and personality/emotional factors should be undertaken to rule out bracketsing conditions.

CHARACTERIZING THE NATURE OF THE DEFECT

In the visual sphere, the recognition of objects, colors, words, geometric forms, faces, and problems and signs should be evaluated. In the event of failure to recognize, the patient should be allowed to match misidentified items to sample and to produce drawings of objects not identified. Quantitative achievement and qualitative performance on these tasks should be carefully noted, keeping in mind that quantitatively correct matching and accurate drawing do not necessarily suggest intact perceptual processing. Poor drawing does not necessarily implicate an apperceptive defect, since visuo-motor or constructive defects may also be present. For this reason, it is important to use tasks toward the end of those which draw from the patient's perception, if possible. Cross-modal matching and matching objects across different views should be evaluated. Line drawings to be copied should contain sufficient internal detail so that slavish tracing of an outline can, if present, be elicited.

Other perceptual functions, such as figure-ground perception (hidden figures), closure and synthetic ability, topographical orientation, route-finding, and visual counting (counting dots on a white paper, picking up pennies spread over a tablet) should also be evaluated. Visual memory for designs, objects, faces, and colors should be assessed by delayed recall (drawing from memory) and multiple-choice recognition tasks. The ability to categorize, sort misidentified objects, and pair similar objects that are not morphologically identical should be tested.

The patient should be asked to identify pictures of well-known people and to identify hospital staff by face. If recognition does not occur, the patient should be asked to determine whether the face is of a male or female or whether the face is of a human or animal. In the acutely hospitalized patient, ability to recognize visiting family can be assessed by dressing the
AGNOSIA

It is now clear that recognition is not a unitary, or even a two-step, process. Much of the historic debate about the existence of agnosia can be attributed to the fact that our models of recognition have generated some incorrect assumptions about what happens when the recognition system becomes damaged. The best example is the debate over whether agnosia is a perceptual or memory access problem. Historically, the major problem with this question has been our assumption that it is the correct question to ask in the first place. We now know better, and, as a result, significant advances have been made in the last decade. The concept of recognition encompasses a broad range of behaviors, including attention, feature extraction, exploratory behavior, pattern and function perception, temporal resolution, and memory. New data on sensory and perceptual systems have revealed the exquisite complexity of the cortical and subcortical systems that support sensory and perceptual activities. Because of these advances, we have transcended the notion of a two-stage recognition model and begun to think in terms of more complex perceptual processes and associations. Instead, recognition of sensory stimuli is now understood as a complex outcome of parallel processing occurring simultaneously at cortical and subcortical levels.

These complexities make it extremely unlikely that a core deficit responsible for all agnostic phenomena exists. It now seems more fruitful to specify the conditions under which stimuli can and cannot be recognized and to more precisely specify the input, processing, and output requirements of specific tasks of identification and recognition. By doing this, and by correlating the emerging clinical findings with available neuropsychological data, a meaningful understanding of the spectrum of agnostic deficits and of normal recognition abilities is rapidly emerging.

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CONCLUSION

During the past decade, significant advances have been made toward an understanding of the complex components of recognition processes. It is important to allow the patient to draw misidentified objects or to after-select them from a group tactually. Tactile exploratory behavior (palpation of objects) should also be carefully observed.
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