THE AGNOSIAS

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The Agnosias are rare disorders in which a patient with brain damage becomes unable to recognize or appreciate the identity or nature of sensory stimuli. Clinical examination of the patient reveals a profound, modality-specific recognition impairment that cannot be fully explained by problems in elementary sensory processing, mental deterioration, attentional disturbances, aphasic misnaming, or to unfamiliarity with the stimuli used to assess recognition abilities. Classically, a distinction between apperceptive and associative forms of agnosia has been made whereby the patient with apperceptive agnosia is said to have deficits in early stages of perceptual processing while the patient with associative agnosia either does not display such problems or does so to a degree not sufficient to substantially impair the ability to perform perceptual operations. The associative agnosic can typically draw, copy, or match unidentified objects, while the apperceptive agnosic cannot. This distinction has been clinically useful, though it is clear that nearly all agnosics have some degree of perceptual (“apperceptive”) disturbance. It should be remembered that adequate copying or matching by itself does not indicate normal perceptual processing (see Farah, 1990; Bauer & Demery, 2003).

**Clinical assessment** of the putative agnosic patient has two fundamental goals. **First,** the possibility that the recognition disturbance exists because of elementary sensory disturbance, dementia, aphasia, or unfamiliarity with the stimulus should be ruled out with standardized neuropsychological testing instruments. **Second,** the scope and nature of the patient’s recognition disturbance should be determined. Does the recognition disturbance exist only for certain stimuli or classes of stimuli? Is it restricted to a particular sensory modality? Under what conditions (if
any) can the patient recognize stimuli? This phase of the evaluation often requires detailed testing using specially formulated testing materials, and should be conducted from the point of view of cognitive models of recognition disturbance (see below). Appropriate referrals for neurologic, neuroradiologic, and basic sensory-perceptual (e.g., ophthalmologic, audiologic) testing are often important in formulating a clinical diagnosis.

I. BASIC DEFINITIONS

Several types of agnosia have been identified in the literature. References summarizing the basic subtypes, clinicoanatomic correlations, and neurobehavioral mechanisms producing disturbances in recognition include Bauer & Demery (2003) and Farah (1990). Humphreys & Riddoch (1987) provide an excellent book-length description of a visual agnosic written from a cognitive neuropsychology perspective. “Pure” forms of these disorders are quite rare and the etiology of the patient’s disorder (e.g., whether from focal stroke vs. a more diffuse cause such as carbon monoxide poisoning) and/or the stage of recovery (if acute onset) will determine the observed pattern of deficits. Defining characteristics of the basic subtypes of agnosia are given below and in Table 1. The remainder of this section provides basic characteristics of agnosia in outline form. This method of presentation is intended to stimulate attempts at differential diagnosis, but should not discourage attempts at more in-depth analysis of presenting syndromes.

I.1. VISUAL AGNOSIAS


a. Key Features

1. cannot recognize the meaning of visually-presented objects

2. disorder is not restricted to naming (e.g., patient cannot point to the object when named or describe or demonstrate its use)
3. sometimes, recognition is better for real objects than for pictures or line drawings

4. can recognize objects when presented in other modalities

b. Varieties:

1. apperceptive: cannot demonstrate adequate perception of object through drawing, copying, or matching tasks

2. associative: drawing, copying, or matching tasks bring more success, though performance is sometimes ‘slavish’

c. Sometimes recognition disturbance is worse for certain categories of objects (e.g., living things, tools, etc.); recognition testing should employ various categories of objects

I.1.2. **Simultanagnosia.**

a. Key Features

1. Patient cannot apprehend the overall meaning of a picture or stimulus, but may be able to appreciate and describe isolated elements.

b. Varieties

1. “dorsal” simultanagnosia (bilateral occipitoparietal lesions); cannot see more than one object at a time

2. “ventral” simultanagnosia (left inferior occipital lesions); may be able to “see” more than one object at a time

c. Often considered a variant of apperceptive agnosia

I.1.3. **Prosopagnosia.**
a. Key Features:
   1. unable to recognize the identity of viewed faces
   2. often can appreciate aspects of faces such as age, gender, or emotional expression.

b. Varieties: Apperceptive and associative forms have been identified on the basis of matching tasks.

c. Associated features: Within-class recognition of other types of visually similar objects (e.g., recognition of individual chairs, cars, animals, etc.) may be impaired

I.1.4. **Color Agnosia.** Because colors can only appreciated visually, the status of ‘color agnosia’ as a true agnostic deficit has been difficult to establish. Nonetheless, four classes of patients have been identified with disproportional impairment in recognizing, naming, or otherwise utilizing color information.

   a. **central achromatopsia:** acquired deficit in color vision due to CNS disease. Cannot match, discriminate, or name colors. Suspect bilateral occipital lesions, but may be unilateral

   b. **color anomia:** specific difficulty in naming colors, usually found in the context of right homonymous hemianopia and pure alexia (Geschwind, 1965). Other aphasic signs generally absent; suspect posterior left hemisphere lesion

   c. **specific color aphasia:** seen in the context of aphasia, represents a disproportionate difficulty in naming colors; suspect left (dominant) parietal lobe damage
d. **color agnosia**: this is a residual category of patients who have difficulty appreciating the nature or name of color they see, but who do not fall within the categories above

I.1.5 **Optic Aphasia**.

a. Key Features:
   1. patient cannot name a visually-presented object
   2. *can* demonstrate its use by gesture, or can point to it when named

b. not regarded as a true agnosia

c. may represent a visual-verbal disconnection

I.2. **AUDITORY AGNOSIAS**

Subtypes of auditory agnosia have been distinguished on the basis of the type of auditory stimulus the patient has difficulty recognizing (Bauer & McDonald, 2003). Although much remains to be understood about these disorders (which have not been studied as exhaustively as cases of visual agnosia), three general classes of deficits have been described.

I.2.1. **Cortical Auditory Disorder and Cortical Deafness**.

a. Key Features:
   1. difficulty recognizing auditory stimuli of many kinds, verbal and nonverbal.
   2. Basic audiologic testing is abnormal

b. Varieties:
   1. cortical deafness: complains of a subjective sense of deafness,
   2. cortical auditory disorder: no subjective sense of deafness
c. Such patients may evolve to one of the more selective types of auditory
agnosia described below; longitudinal assessment is important

**I.2.2. Pure Word Deafness.** (Buchman et al., 1986)

a. Key Features:

1. inability to comprehend spoken language but can read, write, and speak in a relatively normal manner.
2. comprehension of nonverbal sounds is relatively spared.
3. patient is relatively free of aphasic symptoms found with other disorders affecting language comprehension

**I.2.3. Auditory Sound Agnosia (Auditory Agnosia for Nonspeech Sounds).**

a. Key Features

1. inability to comprehend meaning of common environmental sounds, with relative sparing of speech comprehension
2. far more rare than pure word deafness

b. Varieties (Vignolo, 1969):

1. perceptual-discriminative form: makes predominantly acoustic errors (e.g., “whistling” for birdsong)
2. semantic-associative form: makes predominantly semantic errors (e.g., “train” for automobile engine)

**I.2.4. Sensory (Receptive) Amusia.**

a. Key Features

1. inability to appreciate various characteristics of heard music
2. impairment in perceptual vs. conceptual aspects of music should be evaluated

b. impaired music perception occurs to some extent in all cases of auditory sound agnosia, and in most cases of aphasia and pure word deafness; exact prevalence unknown

c. probably underreported because a specific musical disorder rarely interferes with everyday life.

d. perception of pitch, harmony, timbre, intensity and rhythm may be affected to different degrees or in various combinations

I.3. TACTILE AGNOSIAS. Compared to visual agnosias, somatosensory (tactile) agnosias have received scant attention and are poorly understood. Several distinct disorders have been identified, and many classifications of tactile agnosia have been offered. Delay (1935) distinguished three disorders, including: (a) “amorphognosia”, impaired recognition of the size and shape of objects, (b) “ahylognosia”, impaired recognition of the distinctive qualities of objects such as weight, density, texture, and thermal properties, and (c) “tactile asymbolia”, impaired recognition of tactile objects in the absence of amorphognosia or ahylognosia. Although only tentative, a clinically useful distinction can be made between “cortical tactile disorders” (which probably encompass the first two of Delay’s deficit classes), and “tactile agnosia”, which represents an inability to appreciate the nature of tactually manipulated objects.
I.3.1 **Cortical Tactile Disorders.**

a. Key Features

1. deficits in appreciating distinct object qualities such as size, shape, weight, or spatial configuration of tactually presented objects.

b. Varieties: some patients have especially obvious defects of size discrimination, while others fail in tasks which emphasize the spatial character of tactually manipulated objects.

c. No hemispheric specialization exists in elementary somatosensory function, but patients with right hemisphere disease may have difficulty in performing the spatial component of many tactile discrimination tasks.

I.3.2 **Tactile Agnosia.**

a. Key Features:

1. cannot identify objects placed in the hand despite elementary sensory function intact

b. Varieties:

1. Deficit exists in both hands: an “agnosic” deficit (an inability to appreciate the nature of stimuli because of a central defect in processing the nature of a stimulus); cannot demonstrate use of object through gesture
2. Deficit exists in one (usually left) hand: a “visual-verbal disconnection”; can demonstrate use of the object, and can name the object if placed in the other hand

II. Neuroanatomical Correlates

Lesion localization based on individual case studies and recent reviews of the agnosic syndromes described above is presented in Table 2. In general, apperceptive agnosias involve more extensive damage to sensory association cortex while associative agnosias result from lesions of corticocortical pathways or from impairment in those areas where semantic representations of objects are stored. In most published cases, lesions are caused by ischemic stroke, though cases of carbon monoxide poisoning, post-traumatic hematoma, and neoplasm have been reported (Bauer & Demery, 2003; Farah, 1990). It is becoming increasingly recognized (most prominently in the visual domain) that apperceptive agnosia can result from degenerative disease, with particular attention being devoted to dementia syndromes presenting with predominant visuoperceptual disturbance (Biran & Coslett, 2003; Caselli, 2000; Jackson & Owsley, 2003; Mendez, Mendez, Martin, et al., 1990).
III. Differential Diagnosis of Agnosia

A. Basic Decision-Making Process in Differential Diagnosis.

Diagnosis of the agnosias first proceeds by identifying the basic characteristics of the patient’s recognition defect. The process of reaching a tentative initial diagnosis is outlined in flowchart form in Figure 1.

In applying the flowchart, clinicians should remember that “pure” forms of agnosia are not commonly encountered. The first part of the flowchart (Fig. 1a) presents three “streams” representing visual, auditory, and tactile agnosias, respectively, and outlines basic questions which should be asked in making a tentative initial diagnosis. The second part of the flowchart (Fig. 1b) deals specifically with visual agnosias, which are more common and are better understood than their auditory and tactile counterparts.

The flowchart assumes that simple materials for bedside testing are available (or can be manufactured) and that the clinician consults other disciplines in order to further document the extent of neuroanatomic damage and to better characterize sensory/perceptual function. In many cases, the physician and/or the treatment team make such referrals, but it should be remembered that the informed neuropsychologist can serve as a valuable advisor in insuring that appropriate referrals are made. In addition to an extended behaviorally oriented neurological examination, potentially useful referrals include neuroimaging consults (CT/MRI), evoked potential studies, and referrals to ophthalmology, speech pathology/audiology or other professionals for more detailed evaluation of sensory/perceptual and neurobehavioral status. Obviously, referral
decisions should not be made automatically, but should depend on the likely cost-effectiveness of obtaining the requested information.

**B. Neuropsychological Assessment in Differential Diagnosis.**

Once a tentative diagnosis has been reached (or once the clinician has narrowed the differential diagnosis to a subset of possible disorders based on clinical presentation) formal assessment of neuropsychological skills is indicated. As indicated earlier, neuropsychological assessment of the putative agnosic seeks to (a) rule out alternative explanations of the patient’s deficit, and (b) characterize in more precise terms the nature of the patient’s deficit so that its underlying mechanism and its relationship to pathological anatomy can be understood.

1. **Ruling Out Alternative Explanations.**

As suggested earlier, disturbances of “recognition” can occur in a variety of neurological conditions but are considered ‘agnosic’ only if they exist in the relative absence of aphasia, generalized dementia, impaired attentional capacity or other defect that nonspecifically impairs some or all of the information processing steps involved in object recognition. Therefore, one critical aspect of the assessment of the agnostic patient involves assessment of these ‘bracketing’ conditions in order to rule them out as explanations for the recognition defect. A review of available case reports reveals considerable variability in the methods used for this portion of the assessment. Table 3 presents a reasonable strategy for achieving this goal, though it is recognized that many other tests are available for achieving this purpose.

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Table 3 About Here
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In general, patients should receive basic neuropsychological examination designed to
determine general intellectual status, memory function, linguistic competence, and to assess
sensory-perceptual processing. The clinician may wish to perform a comprehensive
neuropsychological battery in order to better understand the patient’s cognitive strengths and
weaknesses, to document ‘baseline’ functioning, or to assist in treatment planning. Assessment
of language ability (naming, auditory comprehension, fluency, repetition, reading, writing, and
praxis) is especially important in understanding the possible role that linguistic factors might
play in the patient’s recognition defect. A comprehensive aphasia battery (e.g., *Boston
Diagnostic Aphasia Examination*, Goodglass & Kaplan, 1983; *Multilingual Aphasia
Examination*, Benton & Hamsher, 1989; *Western Aphasia Battery*, Kertesz, 1982) is useful for
this purpose, though it may be necessary to perform supplementary tests to insure that naming
and recognition are tested in all sensory modalities.

2. **Characterizing the Nature of the Agnosic Deficit**

Once the patient’s general neuropsychological status has been determined, the clinician
will want to perform further testing to more precisely characterize the nature of the patient’s
recognition deficit. At this stage, cognitive neuropsychological models of the perceptual-
recognition process become helpful in guiding the approach to assessment. A representative
model, adapted from Ellis & Young (1988), is presented in Figure 2. Consulting individual case
reports contained in Tables 1 and 2 will also assist in planning an appropriate assessment.

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Figure 2 About Here
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Figure 2 draws on a diverse literature in perceptual psychology and neuropsychology (Ellis & Young, 1988) and is presented to the clinician because such models have succeeded in parsing the process of object recognition into distinct information-processing components or stages. The left side of Figure 2 represents dissociable stages of the object recognition process suggested by clinical and experimental research. The right side of Figure 2 presents the most important implications of the model for clinical assessment, and suggests some commonly available tests that can be utilized in “localizing” the defect at a particular processing level. Defects before the level of the “object recognition unit” can be roughly considered apperceptive in nature, while subsequent deficits correspond to associative forms of agnosia. The model presented in Figure 2 is obviously best suited to evaluating a visual recognition disturbance, but should provide guidance in assessing auditory and tactile agnosia as well. A comprehensive evaluation proceeds by evaluating all levels of the model, even in situations where ‘early’ deficits are found.

**IV. Relevant Laboratory, EEG, and Neuroimaging Correlates**

As a general neuropsychological classification, agnosia is not associated with any definitive pattern of abnormality in laboratory tests. EEG and neuroimaging findings vary with the type of agnosia, as might be anticipated from lesion localization data presented in Table 2. The most common etiologies of agnosia include CVA, tumor, carbon monoxide poisoning, closed head injury, and CNS infection, though as indicated earlier, it is becoming increasingly recognized that some cases of degenerative dementia with primary involvement of posterior cortex can present with prominent signs of (primarily apperceptive) agnosia. Medical findings vary with etiology and localization. Because of these considerations, it can be said that laboratory, EEG, and neuroradiological findings *per se* do not typically play an integral role in differential diagnosis. One exception to this rule is the occasional utility of auditory or visual
evoked potentials as a way of determining whether a defect exists in the sensory projection areas as opposed to the primary sensory or association cortex. Instead, the clinician should rely on behavioral factors and should consider the physical findings as confirmatory.

V. Psychological/Psychiatric Comorbidity

The lesions most likely to produce agnosic defects often spare limbic, paralimbic, or frontal regions that, when damaged, produce primary affective or personality changes. For this reason, specific forms of psychopathology are not obligatory accompaniments of agnosic syndromes. However, secondary emotional reactions to the real-life consequences of agnosia are common. Factors such as unemployment, changes in social life, dependency on others for help in everyday activities (i.e. dressing, transportation, eating), and boredom are seen. These major lifestyle changes may lead to depression or adjustment disorders in some individuals, while others may find adaptive ways to cope. As an excellent example, Humphreys and Riddoch (1987) describe in detail how their patient, John, and his wife both cope with John’s visual agnosia. Their description contains evidence of both adaptive and maladaptive compensations. Although epidemiological studies have yet to be conducted, auditory and tactile agnosias seem less likely to produce major life changes so that is may be that such disorders have less deleterious consequences. Such speculations await definitive research.

Another trait sometimes seen in agnosics is sensory compensation. This is an interesting and as yet unresearched phenomenon reported in the animal literature (Horel & Keating, 1969), in which the agnosic comes to rely on intact sensory modalities (e.g., audition and touch in the case of visual agnosia) in exploratory activity. Whether this represents an attempt to achieve an optimal arousal level through sensory stimulation or an attempt to gain understanding of the world through an intact modality remains to be seen. For example, Bauer’s (1984) patient with
severe visual agnosia listens to music constantly to lessen the boredom of living with the disorder. In our experience, substance abuse is a risk in the chronic period, possibly in response to the reduced stimulation that results from an agnosic deficit, and possibly a result of premorbid factors. It should be emphasized that one problem in understanding psychiatric comorbidity in agnosia is that the relative rarity of these syndromes complicates an analysis of whether such problems are caused or exacerbated by the underlying neurological impairment or whether the appearance of such problems reflects preinjury factors which would have exerted themselves in any event. Such issues await systematic research.

VI. Summary

Agnosia refers to an acquired impairment in the ability to recognize the identity or nature of sensory stimuli. It is a relatively rare disorder that can produce significant everyday impairment. No specific laboratory or neuroradiological marker exists, though orderly anatomic findings have been reported in the literature on visual, auditory, and tactile agnosia that should serve, if present, to raise suspicion about the diagnosis in the individual case. Key symptoms, characteristic neuroradiologic findings, and a general assessment approach based on cognitive neuropsychological models of object recognition were summarized in this chapter. Although significant progress has recently been made, much remains to be learned about these complex disorders, and clinicians are encouraged to take a hypothesis-oriented approach in order to enlarge the available knowledge base.
References


Table 1: Subtypes of Agnosia: Defining Characteristics and Key References

<table>
<thead>
<tr>
<th>Subtype</th>
<th>Affected Stimulus Category</th>
<th>Varieties</th>
<th>Basis for Distinction</th>
<th>Suggested Reference</th>
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<tbody>
<tr>
<td><strong>Visual Agnosias</strong></td>
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<tr>
<td>Visual Object Agnosia</td>
<td>objects</td>
<td>a) apperceptive</td>
<td>a) drawing, matching -</td>
<td>Farah, 1990</td>
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<td></td>
<td></td>
<td>b) associative</td>
<td>b) drawing, matching +</td>
<td>Benson &amp; Greenberg, 1969</td>
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<td></td>
<td></td>
<td>Rubens &amp; Benson, 1971</td>
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<tr>
<td>Simultanagnosia</td>
<td>multiple objects or pictures</td>
<td>a) dorsal</td>
<td>a) cannot see multiple items</td>
<td>Hecaen &amp; Ajuriaguerra, 1954</td>
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<tr>
<td></td>
<td></td>
<td>b) ventral</td>
<td>b) can see multiple items</td>
<td>Kinsbourne &amp; Warrington, 1962</td>
</tr>
<tr>
<td>Prosopagnosia</td>
<td>faces</td>
<td>a) apperceptive</td>
<td>a) match, categorize faces -</td>
<td>DeRenzi et al., 1991</td>
</tr>
<tr>
<td></td>
<td></td>
<td>b) associative</td>
<td>b) match, categorize faces +</td>
<td>Pallis, 1955</td>
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<tr>
<td>Color Agnosia</td>
<td>colors</td>
<td>a) achromatopsia</td>
<td>a) failure of color vision</td>
<td>Damasio et al. 1980</td>
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<td></td>
<td></td>
<td>b) color anomia</td>
<td>b) can succeed at nonverbal color tasks</td>
<td>Geschwind &amp; Fusillo 1966</td>
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<td></td>
<td></td>
<td>c) ‘color aphasia’</td>
<td></td>
<td>Kinsbourne &amp; Warrington, 1964</td>
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<td></td>
<td></td>
<td>d) color agnosia</td>
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<td><strong>Auditory Agnosias</strong></td>
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<tr>
<td>Cortical Deafness and</td>
<td>all sounds</td>
<td>a) cortical deafness</td>
<td>a) subjective deafness?</td>
<td>Vignolo, 1969</td>
</tr>
<tr>
<td>Cortical Auditory</td>
<td></td>
<td>b) agnosia</td>
<td>b) patient claims not to be deaf</td>
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<tr>
<td>Disorder</td>
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<tr>
<td>Pure Word Deafness</td>
<td>speech sounds</td>
<td>a) prephonemic</td>
<td>a) auditory acuity generally impaired</td>
<td>Michel et al., 1980</td>
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<td></td>
<td></td>
<td>b) phonemic</td>
<td>b) disorder of phonemic discrimination</td>
<td>Kanshepolsky, et al., 1973</td>
</tr>
<tr>
<td>Nonverbal auditory</td>
<td>nonspeech sounds</td>
<td>a) perceptual</td>
<td>a) misidentifications primarily acoustic</td>
<td>Buchman et al., 1986</td>
</tr>
<tr>
<td>Agnosia</td>
<td></td>
<td>b) associative</td>
<td>b) misidentifications primarily semantic</td>
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<tr>
<td>Sensory (receptive)</td>
<td>musical sounds</td>
<td></td>
<td>--</td>
<td>Bauer &amp; McDonald, 2003</td>
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<td>amusia</td>
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<td><strong>Tactile Agnosias</strong></td>
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<tr>
<td>Cortical Tactile Disorder</td>
<td>tactually presented objects</td>
<td>a) object-based?</td>
<td>a) fail on object discrimination tasks</td>
<td>Delay, 1935</td>
</tr>
<tr>
<td></td>
<td>and object qualities</td>
<td>b) spatial?</td>
<td>b) fail on tasks requiring spatial discrimin</td>
<td>Caselli, 1991</td>
</tr>
<tr>
<td>Tactile Agnosia</td>
<td>tactually presented objects</td>
<td>a) disconnection</td>
<td>a) unilateral; can demonstrate object use</td>
<td>Corkin, 1978</td>
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<td></td>
<td></td>
<td>b) agnosic</td>
<td>b) bimanual, cannot demonstrate object knowledge</td>
<td>Semmes, 1965</td>
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<td></td>
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<td>Geschwind &amp; Kaplan, 1965</td>
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<td></td>
<td></td>
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<td>Hecaen &amp; David, 1945</td>
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Note: +=function is spared; -=function is impaired
<table>
<thead>
<tr>
<th>Disorder</th>
<th>Lesion Localization</th>
<th>Reference</th>
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<tbody>
<tr>
<td><strong>VISUAL AGNOSIAS</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1) Apperceptive VOA</td>
<td>Diffuse, posterior damage to occipital lobes and surrounding regions</td>
<td>Benson &amp; Greenberg, 1969</td>
</tr>
<tr>
<td>2) Associative VOA</td>
<td>Bilateral: Inferior occipitotemporal</td>
<td>Rubens &amp; Benson, 1971</td>
</tr>
<tr>
<td>3) Simultanagnosia</td>
<td>a) Dorsal: Bilateral parietal and superior occipital</td>
<td>Farah, 1990</td>
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<td></td>
<td>Localized bilaterally to either superior occipital or inferior parietal lobes</td>
<td>Hecaen &amp; Ajuriaguerra, 1954</td>
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<td></td>
<td>b) Ventral: Dominant occipitotemporal junction</td>
<td>Kinsbourne &amp; Warrington, 1962</td>
</tr>
<tr>
<td>4) Prosopagnosia</td>
<td></td>
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<tr>
<td>a) Apperceptive</td>
<td>Traditionally seen as bilateral in all or nearly all cases. Cortex and white matter in occipitotemporal gyrus or projection system.</td>
<td>Bauer &amp; Demery, 2003</td>
</tr>
<tr>
<td></td>
<td>More recently a few cases of what appears to be unilateral damage to right visual association cortices within occipital and parietal lobes.</td>
<td>Damasio et al., 1990 DeRenzi, 1986</td>
</tr>
<tr>
<td>b) Associative</td>
<td>Bilateral anterior temporal regions compromising hippocampal and other regions</td>
<td>Damasio et al., 1990</td>
</tr>
<tr>
<td>5) Color Agnosia</td>
<td></td>
<td></td>
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<tr>
<td>a) Achromatopsia</td>
<td>Unilateral or bilateral inferior ventromedial region of occipital lobe - involves lingual and fusiform gyri - superior field defects</td>
<td>Damasio et al., 1982</td>
</tr>
<tr>
<td>b) Color Anomia</td>
<td>Dominant occipital infarction with corpus callosum involvement</td>
<td>Geschwind &amp; Fusillo, 1966</td>
</tr>
<tr>
<td>c) Specific Color Aphasia</td>
<td>Dominant parietal damage coincident with posterior aphasia</td>
<td>Kinsbourne and Warrington, 1964</td>
</tr>
<tr>
<td>6) Optic Aphasia</td>
<td>Unilateral: Dominant occipital lobe and splenium</td>
<td>Riddoch &amp; Humphreys, 1987</td>
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<td></td>
<td></td>
<td>Geschwind, 1965</td>
</tr>
<tr>
<td><strong>AUDITORY AGNOSIA</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1) Cortical Auditory Disorder</td>
<td>Variable- can involve superior temporal gyrus and efferent connections of Heschl’s gyrus or bilateral subcortical lesions</td>
<td>Kazui et al., 1990</td>
</tr>
<tr>
<td></td>
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<td>Oppenheimer and Newcombe, 1978</td>
</tr>
<tr>
<td>2) Pure Word Deafness</td>
<td>Bilateral: Symmetrical lesions of anterior section of superior temporal gyri - Most often bilateral disconnections of Wernicke’s area from auditory input</td>
<td>Buchman, et al., 1986</td>
</tr>
<tr>
<td></td>
<td>Unilateral (Rare): Deep subcortical in dominant superior temporal region damaging primarily auditory cortex and/or pathways to and from medial geniculate gyrus</td>
<td>Weisenburg and McBride, 1935/1964</td>
</tr>
<tr>
<td>3) Auditory Sound Agnosia</td>
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</tr>
<tr>
<td>a) Perceptual-Discrimination</td>
<td>Nondominant hemisphere</td>
<td>Vignolo, 1969</td>
</tr>
<tr>
<td>Type</td>
<td>Dominant hemisphere - linked with posterior aphasia</td>
<td>Vignolo, 1969</td>
</tr>
<tr>
<td>------</td>
<td>---------------------------------------------------</td>
<td>---------------</td>
</tr>
<tr>
<td>4) Sensory (Receptive) Amusia</td>
<td>Unilateral temporal lobe - if comorbid with aphasia, lesion is on dominant side.</td>
<td>Bauer and McDonald, 2003</td>
</tr>
</tbody>
</table>

**TACTILE AGNOSIAS**

<table>
<thead>
<tr>
<th>Type</th>
<th>Severe and Long-Lasting: Contralateral postcentral gyrus Less severe, bilateral lesions of SII</th>
<th>Corkin, 1978</th>
</tr>
</thead>
<tbody>
<tr>
<td>1) Cortical Tactile Disorders</td>
<td>Corpus callosum (affecting crossing somatosensory fibers (minimally; actual lesion may be more extensive)</td>
<td>Geschwind &amp; Kaplan, 1962</td>
</tr>
<tr>
<td>2) Unilateral Tactile Anomia</td>
<td>Contralateral primary somatosensory projection area in postcentral gyrus</td>
<td>Caselli, 1991</td>
</tr>
</tbody>
</table>
Table 3: Ruling out Alternative Causes of Recognition Disturbance

<table>
<thead>
<tr>
<th>Condition or Problem</th>
<th>Assessment Instruments</th>
<th>Domains Tested</th>
<th>Reference</th>
</tr>
</thead>
<tbody>
<tr>
<td>Generalized Dementia</td>
<td>Dementia Rating Scale</td>
<td>memory, attention/concentration, construction, initiation/perseveration</td>
<td>Mattis (1988)</td>
</tr>
<tr>
<td>Aphasia</td>
<td>Boston Diagnostic Aphasia Exam</td>
<td>fluency, comprehension, naming, repetition, reading, writing, praxis</td>
<td>Goodglass &amp; Kaplan (1983)</td>
</tr>
<tr>
<td></td>
<td>Multilingual Aphasia Exam</td>
<td></td>
<td>Benton &amp; Hamsher (1989)</td>
</tr>
<tr>
<td></td>
<td>Western Aphasia Battery</td>
<td></td>
<td>Kertesz (1982)</td>
</tr>
<tr>
<td>Disturbances of Attention/Orientation (e.g.,</td>
<td>Temporal Orientation Test</td>
<td>time orientation</td>
<td>Benton, Sivan, et al. (1994)</td>
</tr>
<tr>
<td>delirium)</td>
<td>Visual Search and Attention Test</td>
<td>visual search and selectivity</td>
<td>Trenerry et al. (1990)</td>
</tr>
<tr>
<td></td>
<td>WAIS-R Digit Span</td>
<td>focused attention span</td>
<td>Benton &amp; Hamsher (1989)</td>
</tr>
<tr>
<td></td>
<td>Sentence Repetition</td>
<td>focused attention span (sentences)</td>
<td>Schenkenberg et al. (1980)</td>
</tr>
<tr>
<td></td>
<td>WMS-R Mental Control</td>
<td>mental tracking, sustained attention spatial attention, hemispatial neglect</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Line Bisection</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Unfamiliarity with Stimuli</td>
<td>Determined subjectively; the examiner needs to insure that</td>
<td>visual, auditory, and tactile object identification with common objects</td>
<td>Familiarity must be determined,</td>
</tr>
<tr>
<td></td>
<td>failures of naming/identification are not based on</td>
<td>should be tested in each patient to determine familiarity statistics and to</td>
<td>even informally, on an</td>
</tr>
<tr>
<td></td>
<td>experiential, cultural, or other factors that lead to the</td>
<td>determine modality specificity; subjects who cannot name objects should be</td>
<td>individual-subject basis. If</td>
</tr>
<tr>
<td></td>
<td>patient’s unfamiliarity with stimuli tested; use of common</td>
<td>encouraged to divulge anything they know about it or to group items into</td>
<td>creating in-house stimulus</td>
</tr>
<tr>
<td></td>
<td>or frequently-encountered items</td>
<td>familiar and unfamiliar categories</td>
<td>sets, general references</td>
</tr>
<tr>
<td></td>
<td>typically circumvents this problem</td>
<td></td>
<td>containing relevant statistics</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>on item frequency, imageability,</td>
</tr>
</tbody>
</table>
Figure 1a
Flowchart for Clinical Decision-Making
Differential Diagnosis of Agnosia

Patient with Recognition Disturbance

Is disorder modality-specific?

No

Evaluate for dementia, aphasia, acute confusional state, etc.

Yes

Visual Recognition Impaired in excess of other modalities

Can patient demonstrate the use of objects or point to them when named?

Yes

OPTIC APHASIA

No

WORK-UP FOR APHASIA

Auditory Recognition Impaired in excess of other modalities

Does patient have signs of posterior aphasia (e.g., are there abnormalities in speaking, writing, and reading)?

Yes

APPerceptive Visual Agnosia

No

non-speech > speech

ASSOCIATIVE VISUAL AGNOSIA

Tactile Recognition Impaired in excess of other modalities

Can patient discriminate object characteristics (shape, weight, thermal properties)?

Yes

AUDITORY SOUND AGNOSIA

No, or minor

SENSORY (RECEPTIVE) AMUSIA

Yes

TACTILE-VERBAL DISCONNECTION (UNILATERAL TACTILE ANOMIA)

No

TACTILE AGNOSIA

OPTIC APHASIA

Does patient have severe disturbance in acuity, field, visual attention, or other primary visual ability?

Yes

PURE WORD DEAFNESS

No

speech > nonspeech

AUDITORY SOUND AGNOSIA

What sounds are affected?

speech > nonspeech

non-speech > speech

LIMITED TO ONE HAND

both

music only

LIMITED TO ONE HAND

Bilateral

Is disorder limited to one (usually the left) hand, or is it bilateral?
Figure 1b: Differential Diagnosis of Visual Agnosia

1. Patient with Visual Recognition Disturbance (determined by screening or primary complaint)
   \[\rightarrow\] Specific to Vision?
   \[\rightarrow\] No
   \[\rightarrow\] Consider other Disorders
   \[\rightarrow\] Yes

   What class of stimuli is most affected? (NOTE: Patient may qualify for more than one branch [e.g., may have impairments in recognizing both objects and faces]; follow all that apply)
   \[\rightarrow\] Objects
   \[\rightarrow\] Faces
   \[\rightarrow\] Colors
   \[\rightarrow\] Patient seems to have difficulties with a variety of complex stimuli

   Does patient have severe disturbance in acuity, field, visual attention, or other primary visual ability? Or, if screening, does the patient fail in copying or matching misidentified stimuli?
   \[\rightarrow\] Yes
   \[\rightarrow\] No

   Does the patient display a primary defect of color vision or complain of dull, “washed-out” vision? (Formal testing with color perception tasks may be needed to answer this question)
   \[\rightarrow\] Yes
   \[\rightarrow\] No

   Is the patient impaired in discriminating or matching unrecognized faces?
   \[\rightarrow\] Yes
   \[\rightarrow\] No

   Is the color naming deficit limited to the visual modality, or does the patient fail at all color-related tasks (e.g., succeeds at dot counting)
   \[\rightarrow\] Limited to vision
   \[\rightarrow\] Fails all

   Does the patient appear capable of seeing more than one item at a time (e.g., succeeds at dot counting)
   \[\rightarrow\] Yes
   \[\rightarrow\] No

   Is the patient impaired in discriminating or matching unrecognized faces?
   \[\rightarrow\] Yes
   \[\rightarrow\] No

   Does the patient have severe disturbance in acuity, field, visual attention, or other primary visual ability? Or, if screening, does the patient fail in copying or matching misidentified stimuli?
   \[\rightarrow\] Yes
   \[\rightarrow\] No

   Does the patient display a primary defect of color vision or complain of dull, “washed-out” vision? (Formal testing with color perception tasks may be needed to answer this question)
   \[\rightarrow\] Yes
   \[\rightarrow\] No

   Is the patient impaired in discriminating or matching unrecognized faces?
   \[\rightarrow\] Yes
   \[\rightarrow\] No

   Is the color naming deficit limited to the visual modality, or does the patient fail at all color-related tasks (e.g., succeeds at dot counting)
   \[\rightarrow\] Limited to vision
   \[\rightarrow\] Fails all

   Does the patient appear capable of seeing more than one item at a time (e.g., succeeds at dot counting)
   \[\rightarrow\] Yes
   \[\rightarrow\] No
Figure 2
Clinical Application of Cognitive Neuropsychological Model

Viewed object

- Initial Representation
  (basic form and shape)

  | Key issue: can subject appreciate basic object form and shape qualities? Object and shape discriminations (e.g., Warrington & James, 1991), Visual Closure (e.g., Street, 1931), Visual Form Discrimination (Benton et al., 1994)

- Viewer-Centered Representation
  (object qualities and features from viewer’s perspective)

  | Key issue: can subject match identical objects, or discriminate between same and different items? Birmingham Object Recognition Battery (Riddoch & Humphreys, 1993), Benton Face Recognition, items 1-6 (Benton et al., 1994)

- Object-Centered Representation
  (object qualities and features independent of view)

  | Key issue: can subject match objects presented in different views? Birmingham Object Recognition Battery (Riddoch & Humphreys, 1993); Benton Face Recognition, items 7-22; (Benton et al., 1994)

- Object Recognition Units
  (stored representation of familiar object)

  | Key issue: is the item familiar or not? Birmingham Object Recognition Battery (Riddoch & Humphreys, 1993); familiarity discriminations; famous face recognition from Albert Remote Memory Battery (Albert et al., 1979 and subsequent modifications)

- Semantic System
  (meaning information about objects; information about object class or about the nature of individual item)

  | Key issue: can subject recognize the general class to which item belongs? Have subject group like objects together (e.g., tools, office items) on the basis of semantic similarity; Birmingham Object Recognition Battery (Riddoch & Humphreys, 1993)

- Name Retrieval
  (object names)

  | Key issue: can subject derive the specific name for the presented item? Confrontation naming tests (Benton et al., 1989; Kaplan et al., 1983)

  spoken name