Chapter 9

Disorders of higher visual processing

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Beyond the striate cortex, visual information is distributed among a wide array of cortical regions for more specialized forms of processing. Neuropsychological studies of subjects have made important contributions to our understanding of the anatomical substrate of visual processing, complementing neurophysiological experiments in monkeys and, more recently, functional neuroimaging work in humans.

Although the number of cortical regions involved in vision is large, conceptually it has been useful to group both processes and their disorders into two main groups (Ungerleider and Mishkin, 1982). A ventral pathway based on medial occipitotemporal structures appears to contain modules critical for object recognition and the basic processing of form and color that is required. A dorsal pathway based on lateral occipitoparietal structures is involved in motion processing and spatial processes such as attention and localization. Colloquially these have been dubbed the "what" and "where" pathways, though a competing formulation has suggested that the dorsal pathway is configured for preparing responses to the environment, an "action" pathway (Milner and Goodale, 1995). In this chapter we follow this useful division, considering disorders of color processing and object recognition under the heading of ventral pathway disorders, and then disorders of motion processing and spatial processing as dorsal pathway disorders. Last, we finish with a consideration of the phenomenon of blindsight, which focuses the aspects of vision these regions and subcortical structures can support in the absence of the striate cortex.

VENTRAL OCCIPITOTEMPORAL PATHWAY DISORDERS

Disorders of color processing

CEREBRAL (CENTRAL) DYSCHROMATOPSIA/ ACHROMATOPSIA

Cerebral dyschromatopsia is impaired color perception due to an acquired brain lesion. In achromatopsia there is complete absence of color perception. Subjects with achromatopsia complain that everything appears in shades of gray (MacKay and Dunlop, 1899; Pallis, 1955; Meadows, 1974a; Rizzo et al., 1993), sometimes less bright (Rizzo et al., 1993) or tinged a "dirty gray" (Damasio et al., 1980). Less frequently vision is tinted, as if peering through a colored filter (Critchley, 1965; Koyama et al., 2006). Impaired color discrimination can affect a number of daily activities, such as distinguishing money, stamps, and traffic lights (Sacks, 1995).

Hemiachromatopsia is loss of color limited to the contralateral hemifield. Typically it is asymptomatic until the defect is demonstrated (Albert et al., 1975; Paulson et al., 1994). Hemiachromatopsia is usually associated with a homonymous superior quadrantanopia (Albert et al., 1975; Damasio et al., 1980; Paulson et al., 1994), and so the color defect is only demonstrable in the lower quadrant. Two rare cases with color defects limited to one quadrant have been described (Kölmel, 1988): it is not clear whether these were true chromatic defects or subtle relative scotomata, but the quadrantic representation of the human V4 area on functional neuroimaging (Wandell and Wade, 2003) suggests that a quadrantic dyschromatopsia is theoretically possible.

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The incidence of hemiachromatopsia is underestimated, given that it is asymptomatic and not detected by routine clinical tests.

On testing, achromatopsic subjects cannot name colors, though those with a partial defect may be able to name broad color categories like red or yellow. Color discrimination tests are a better assessment than color naming. One can begin with pseudoisochromatic plates (Hardy et al., 1957; Ichikawa et al., 1987) but caution is required as some achromatopsic subjects can see the numbers if the plates are so far away that the individual dots merge, allowing color boundaries to emerge (Meadows, 1974a; Ichikawa et al., 1987; Victor et al., 1989; Heywood et al., 1991). The Nagel anomaloscope asks observers to find the correct mix of yellow-green and yellow-red lights to match a yellow monochromatic light: healthy subjects find a unique solution but achromatopsic subjects accept a wide range of mixtures as a match (Pearlman et al., 1979; Rizzo et al., 1993). The best tests for achromatopsia require the subject to sort color chips. Hue discrimination (e.g., red versus green) is tested by the Farnsworth-Munsell 100 hue test or the shorter D-15 test. The Sahlgren saturation test (Frisén and Kalm, 1981) assesses saturation perception (e.g., pink versus red) and the lightness discrimination test assesses brightness perception, with chips of dark to light gray (Verriest et al., 1979; Pinckers and Verriest, 1987). Achromatopsic subjects often have abnormal discrimination of hues and saturation but normal perception of brightness (Heywood et al., 1987, 1991; Victor et al., 1989; Rizzo et al., 1993). Unlike the defect in congenital color blindness, cerebral achromatopsia affects perception of all hues, although not necessarily equally (Rizzo et al., 1993). Besides these tests, assessment of the duration and hue of color afterimages can also reveal deficits after problems with hue discrimination have resolved (Koyama et al., 2006).

Hemiachromatopsia can be demonstrated by moving a colored object from the contralateral to the ipsilateral hemifield: the subject will note a sudden appearance of color on what had been a gray object when it crosses the vertical meridian. More detailed testing of hemiachromatopsia is difficult, as many of the standard tests are designed for central viewing and can still be performed well when color perception is intact in at least one hemifield (Rizzo et al., 1993).

Achromatopsia may also affect color constancy. The wavelengths projecting from an object to the eye depend upon both its reflectant properties and the illumination; nevertheless, perceived object color is stable under a wide range of lighting conditions (Land, 1986; Zeki, 1990). An apple looks red whether in sunlight or under fluorescent light, in an orchard or in a grocery display. "Discounting the illuminant" requires neural

computations in retina and cortex that average the spectral composition from large regions of the background to infer the illumination, which is then taken into account to judge true object color (Land et al., 1983; Land, 1986). Failure to perform this integration or 'anchoring' will result in poor color constancy, with color percepts that vary with changes in lighting. One cannot determine this in achromatopsic subjects, who have no color percept at all, but studies have shown impaired color constancy in dyschromatopsic subjects, who have some residual hue sensitivity (Kennard et al., 1995; Clarke et al., 1998; D'Zmura et al., 1998; Hurlbert et al., 1998; Kentridge et al., 2004).

Subcortical contributions to color processing from cones and parvocellular retinal ganglion cells can still be discerned in cerebral achromatopsia. Photopic spectral sensitivity curves (Heywood et al., 1991, 1996; Kennard et al., 1995) and evoked potential or psychophysical measures of chromatic contrast sensitivity (Heywood et al., 1996; Adachi-Usami et al., 1997) show residual evidence of trichromacy and color opponency. Anomaloscopic testing shows that achromatopsia resembles anomalous trichromacy rather than monochromacy, despite the subjective report of vision as monochromatic "shades of gray" (Pearlman et al., 1979). Achromatopsics can also use color-opponent signals to detect a difference between colors and locate boundaries between differently colored regions, even though they do not know what the colors are (Kentridge et al., 2004). This local chromatic contrast can support the perception of color-defined form or the movement of chromatic stimuli (Cavanagh et al., 1998; Heywood et al., 1998; Cole et al., 2003), and pupillary responses to color (Cowey et al., 2008a). In distinction, local chromatic contrast is lost, not with achromatopsia, but following striate lesions (Kentridge et al., 2007).

Achromatopsia is caused by bilateral lesions of the lingual and fusiform gyri (Verrey, 1888; Zeki, 1990), as evident on neuroimaging (Green and Lessell, 1977; Pearlman et al., 1979; Damasio et al., 1980; Victor et al., 1989; Heywood et al., 1991; Rizzo et al., 1993). Hemiachromatopsia occurs with unilateral right- or left-sided lesions (Freedman and Costa, 1992; Short and Graff-Radford, 2001) (Fig. 9.1). Lesions of the middle third of the lingual gyrus or the white matter behind the posterior tip of the lateral ventricle are critical (Damasio and Frank, 1992; Rizzo et al., 1993; Bouvier and Engel, 2006). Initially it was thought that these lesions might damage a human homolog of area V4, which was the first cortical region found to have color-selective responses (Zeki, 1990). However, impaired hue perception in monkeys occurs not with lesions of V4 (Dean, 1979; Wild et al., 1985; Heywood and Cowey, 1987; Heywood et al., 1992; Walsh et al.,



Fig. 9.1. Hemiachromatopsia. Magnetic resonance axial image from a 77-year-old woman with a stroke (arrow indicates its hyperintense signal) affecting the lingual gyrus in the left posterior cerebral arterial territory, causing right upper quadrantanopia with hemiachromatopsia apparent in the right lower quadrant.

1992; Schiller, 1993) but with extensive bilateral lesions that include areas TE and TEO (Heywood et al., 1995; Cowey et al., 2001). In humans, functional neuroimaging reveals several color-processing regions, including a V4 homolog, a second area named V4alpha or V8 in the fusiform gyrus (Hadjikhani et al., 1998; Bartels and Zeki, 2000), and more distant regions (Gulyás and Roland, 1991; Gulyás et al., 1994; Beauchamp et al., 1999). A severe achromatopsic defect may require damage to or disconnection of several components of this color network, and not just a single region (Heywood et al., 1992; Merigan, 1993; Wandell and Wade, 2003). Indeed, functional neuroimaging suggests that the severity of the dyschromatopsic defect may depend upon whether one or several color-processing regions are affected (Beauchamp et al., 2000).

Achromatopsia is often part of a tetrad that includes prosopagnosia, topographagnosia, and superior homonymous field defects. Less frequently associated deficits include general visual agnosia (Heywood et al., 1991; Ogden, 1993), alexia when there is a right hemianopia (Meadows, 1974a; Green and Lessell, 1977), and amnesia when damage extends to the anterior temporal lobe (Meadows, 1974a; Ogden, 1993). Following research on the effects of V4 lesions in monkeys, more detailed testing in some subjects has shown impaired detection of stimuli with low salience (Mendola and Corkin, 1999), indicating inefficient allocation of attention in form processing.

Achromatopsia is most often due to strokes, such as bilateral sequential or simultaneous infarctions from

posterior cerebral arterial occlusions or a coagulopathy (Orrell et al., 1995). Achromatopsia may be the initial symptom or the final outcome from a resolving cortical blindness. Other bilateral lesions causing achromatopsia include herpes simplex encephalitis (Heywood et al., 1991), cerebral metastases (Green and Lessell, 1977), repeated focal seizures (Aldrich et al., 1989), focal dementia (Freedman and Costa, 1992), and even migraine aura (Lawden and Cleland, 1993). Temporooccipital white-matter damage has caused a reversible dyschromatopsia in one subject with carbon monoxide poisoning (Fine and Parker, 1996).

COLOR ANOMIA AND AGNOSIA

While achromatopsic subjects cannot discriminate hue and saturation, though some dyschromatopsic subjects can name some colors, the opposite is true of subjects with either color anomia or color agnosia. These subjects can discriminate colors accurately but cannot name them, and may not be aware of this problem.

Color anomia may occur as part of a more general anomic aphasia or as a specific entity. The latter occurs with left occipital lesions and often with a right homonymous hemianopia, in contrast to the superior field loss typical in cerebral achromatopsia. Color anomia may be due to an interhemispheric visual-verbal disconnection when it is associated with pure alexia. Loss of connections in the splenium prevents color information from the intact left hemifield and right striate cortex from accessing language processors in the left angular gyrus (Holmes, 1950; Geschwind and Fusillo, 1966; Oxbury et al., 1969; de Vreese, 1991). Some, but not all, of these subjects can name visual objects, which has been explained by speculation that, unlike colors, objects can activate not only visual but also somasthetic representations of object shape, whose information is transferred in more anterior parts of the corpus callosum (Geschwind and Fusillo, 1966).

Subjects with color dysphasia have trouble not only with naming colors they see, but also with naming the colors of familiar objects that are imagined or depicted in grayscale, tasks that subjects with the disconnection form of color anomia can do well (Oxbury et al., 1969). This suggests loss of an internal lexicon for colors. Most have lesions of the left angular gyrus and hence additional deficits such as alexia with agraphia, Gerstmann syndrome, and right homonymous hemifield defects.

Color agnosia is unusual (Kinsbourne and Warrington, 1964; Luzzatti and Davidoff, 1994; Miceli et al., 2001). As in color anomia, these subjects can sort and match colors, and some can even name the colors they see. However, they cannot color line drawings correctly or state whether drawings have been colored correctly. They cannot learn paired associations between seen objects and seen colors (a visual-visual task) but, in contrast to subjects with color amnesia, they can remember the stereotyped colors linked to certain objects, like the red of a fire engine or the yellow of a banana (van Zandvoort et al., 2007). Their knowledge of color categories is also impaired (van Zandvoort et al., 2007). Associated defects include pure alexia, object anomia, and poor object imagery. Most of these subjects have left occipitotemporal lesions, some with a right hemianopia. There is a rare developmental form without visible lesions on imaging, which may be inherited in an autosomal-dominant fashion (Nijboer et al., 2007).

Disorders of object recognition

Impaired visual object recognition is the prototypical functional disorder of lesions of the ventral occipitotemporal pathway. This can vary from forms in which even rudimentary distinctions between forms and shapes have been affected, to more subtle disturbances that are specific for only certain classes of objects. General visual agnosia is the condition typified by the man who mistook his wife for a hat. More selective agnosias are probably more common though still rare, the most prominent being agnosia for words (alexia) and agnosia for faces (prosopagnosia).

GENERAL VISUAL AGNOSIA

Subjects with visual agnosia no longer recognize previously familiar objects and cannot learn to identify new objects by sight alone (Farah, 1990; Riddoch and Humphreys, 2003). A historic debate centers on the necessary and sufficient impairments that generate agnosia, and the extent to which these impairments involve memory rather than perception. Milner and Teuber (1968) defined agnosia as an associative disorder in which percepts are stripped of their meanings. This associative agnosia can be considered a selective disturbance of visual memory. In contrast, perceptual dysfunction is the main cause of disordered visual recognition in apperceptive agnosia (Lissauer, 1890). While useful, it is probable that this apperceptive/associative distinction is rarely encountered in a pure form, though in a particular subject one disturbance may dominate.

In the past, distinctions between apperceptive and associative agnosia were often based on two observations: whether subjects could copy drawings accurately and whether they could match basic shapes. Intact skills on these two tasks would be taken as a sign of adequate perception. However, others have pointed out a) that the intact drawing of some of these subjects was accomplished by a very anomalous and laborious piecemeal strategy, suggesting abnormal perceptual processing after all, and b) there are other reasons beside visual agnosia that may cause subjects to draw poorly (Humphreys et al., 1994). Likewise, slow, effortful, though ultimately correct matching of shapes may not prove integrity of perceptual processing.

More recent work has also examined the nature of the perceptual and associative defects that occur in visual agnosia, and suggested that these two broad categories may be fractionated further, given the multiplicity of processes involved in the complex process of object recognition (Humphreys et al., 1994). In terms of object perception, processes such as shape coding, figure–ground segmentation, the grouping and integration of features into whole objects, and the mapping of the resulting perceptual descriptions to stored structural representations of objects are all important processes, and potentially dissociable from each other.

One influential taxonomy of agnosia has been proposed (Humphreys and Riddoch, 1987; Farah, 1988). This includes several varieties of apperceptive agnosia. "Shape agnosia" or "visual form agnosia" refers to those subjects who have the classical impairment of matching shapes, implying a defect in representing elementary properties of curvature, surface, and volume (Humphreys and Riddoch, 1987; Farah, 2004). Classical examples include subjects Mr S (Efron, 1969) and DF (Milner et al., 1991). Such subjects perform the standard tests of shape matching and the copying of drawings poorly. Shape misperception can fall along a continuum, with some subjects, like SMK, perceiving simple shapes better than more complex ones (Davidoff and Warrington, 1993). Their residual object recognition is fragmentary and often relies on inferences from texture and color. Some benefit from tracing forms with their hands, in effect translating a visual percept into a kinesthetic one (Adler, 1944; Landis et al., 1982).

Types of form information that depend upon cues processed by the dorsal stream may be preserved in visual form agnosia. These subjects can see the form of moving objects when the movement of the object differs from the background. Their drawing of real objects is better when they are allowed to move their head, suggesting that they can perceive form from depth cues, another putative function of the dorsal stream (Chainey and Humphreys, 2001). Also, form information may be available for functions performed by the dorsal stream. Even though they have trouble reporting the orientation of line segments, subjects with visual agnosia can orient their grasp and reach correctly to the orientation of linear objects (Goodale et al., 1991; James et al., 2003). Some can also recognize gestures and the depiction of actions in line drawings, implying a dissociation

between perception of actions and objects (Ferreira et al., 1998).

The pathophysiology of visual form agnosia continues to be debated. One concept is that these subjects have a peppering of minute scotomata across their visual fields from diffuse occipital damage (Campion and Latto, 1985), an explanation that is appealing given the frequent association with carbon monoxide poisoning. However, simulations in healthy subjects have produced mixed results (Vecera and Gilds, 1997, 1998; Abrams and Law, 2002), with a suggestion that agnosic performance is more closely mimicked when the experimenters eliminate grouping cues that can be used to define an object's shape (Vecera and Gilds, 1998). This is consistent with observations that agnosic subjects have problems with such grouping cues (Behrmann and Kimchi, 2003).

Problems with grouping merge into a second form of apperceptive agnosia. Intact ability to perceive elementary shape features but failure to integrate them into a perceptual whole is considered an "integrative agnosia," as in subject HJA (Riddoch and Humphreys, 1987b). These subjects may be able to match simple forms and shapes slowly and slavishly copy drawings in a piecemeal fashion, but have trouble particularly with perceptually constructing items from multiple elements (Humphreys and Riddoch, 1987; Shelton et al., 1994). Appreciation of overall global shape may be intact, without appreciating the elements integrated to form it. This can create problems with recognizing impossible objects (like some Escher drawings), since these require appreciation that the local elements, while plausible in themselves, do not integrate into a correct three-dimensional representation (Delvenne et al., 2004). For similar reasons discriminating real objects from incorrect objects made from parts of other objects can be difficult. These subjects also have trouble disentangling overlapping figures (Riddoch and Humphreys, 1987b; Grossman et al., 1997).

"Transformation agnosia" refers to a more unusual condition in which subjects cannot recognize objects seen from unusual (noncanonical) viewpoints. This suggests a difficulty with deriving a viewpointindependent representation of three-dimensional structure (Warrington and James, 1986). These subjects may not present with typical agnosic complaints of difficulty recognizing objects in daily life, however.

There may also be different subtypes of the associative form of visual agnosia. One consideration is whether the disorder reflects an inability to access intact memories or stored representations of objects (semantic access agnosia) or whether the memory representations have been destroyed (semantic agnosia) (Humphreys and Riddoch, 1987). The distinction can be made by probing with verbal tests what subjects remember about objects, though it has been debated whether a semantic agnosia can be specific for vision, or whether it only ever occurs as part of a multimodal semantic dementia (Farah, 1988). One can also fractionate object knowledge into two broad types: stored structural descriptions, about the shape of the object, and stored semantic knowledge, about their function, location, habitat, or history (Riddoch and Humphreys, 1987a). There are agnosic subjects who can provide accurate descriptions from memory of what objects look like, but cannot name or pantomime their use. They cannot categorize visual objects by semantic similarity (e.g., "is a hammer more similar to pliers or to a comb?"), though they can do so when presented with the names of these objects (Riddoch and Humphreys, 1987a; Carlesimo et al., 1998). Hence these have a defect specific to stored semantic knowledge.

Potentially related to this fractionation of object knowledge is a distinction between living and nonliving things (Caramazza and Shelton, 1998). This is most frequently reported as a semantic distinction, and hence seen mainly in associative agnosia, though some argue that this need not necessarily be the case (Thomas and Forde, 2006). Most frequently reported is a disproportionate impairment for recognition of living things (Farah et al., 1991; Kurbat, 1997). There are many proposed reasons for this dissociation. The obvious one is separate modular representations for living and nonliving things (Kurbat and Farah, 1998). Others suggest that living things are distinguished primarily by their structural properties, whereas nonliving things are coded primarily by their semantic properties, in particular their functionality (Warrington and Shallice, 1984). In a related fashion some argue that, because the most important information about nonliving things is how they are manipulated and used, these objects can access sensorimotor representations that are not relevant for living things (Wolk et al., 2005). Other reasons advanced for the vulnerability of recognition of living things include greater reliance for the recognition of animate objects on global processing (Thomas and Forde, 2006), and greater similarity in perceptual structure between different living things than between different nonliving things, making discriminations harder for the former, which has been confirmed by reaction time data in healthy subjects (Humphreys, 1988).

While some affirm that associative visual object agnosia is a real entity (Riddoch and Humphreys, 2003), its existence has been challenged by others, on the grounds that there is no evidence for separate perceptual and mnemonic representations, or for truly intact perception in a subject purported to have associative visual agnosia (Farah, 2004; Delvenne et al., 2004). It has been argued that some cases considered to have associative agnosia were actually cases of integrative agnosia, in whom only shape or visual form agnosia had been excluded.

The fractionation of visual agnosia into different subtypes is paralleled by the variation in their structural correlates. Visual form agnosia is frequently associated with widespread occipital lesions incurred from diffuse insults, most typically poisoning with carbon monoxide (Adler, 1950; Benson and Greenberg, 1969; Campion and Latto, 1985), in one case with mercury (Landis et al., 1982), and sometimes from bilateral hypoxicischemic occipital injury or posterior cortical atrophy (Fig. 9.2). The cases with integrative agnosia have had bilateral peristriate occipital infarcts or a posterior variant of Alzheimer's disease (Riddoch and Humphreys, 1987b; Grossman et al., 1997). Difficulty with figure segmentation and matching objects across different views may occur with more discrete right occipital damage (Humphreys et al., 1994), as with transformation agnosia (Warrington and James, 1986). Associative agnosia and semantic access deficits may be more prominent with left occipital damage, as with posterior cerebral arterial infarcts (Capitani et al., 2009), frequently involving parahippocampal, fusiform, and lingual gyri (Feinberg et al., 1994), though some claim the necessity of bilateral lesions. In Alzheimer's disease, neurofibrillary tangles in Brodmann areas 18, 19, and 37 are correlated with associative deficits in object recognition, but not with apperceptive deficits as tested with overlapping and hidden figures (Giannakopoulos et al., 1999).

DISORDERS OF FACE PROCESSING

Prosopagnosia

Prosopagnosia is the impaired ability to recognize familiar faces or to learn to recognize new faces (Bodamer, 1947; Barton, 2003). As a symptom, this can occur with more general problems of perception, cognition, and memory, as in macular degeneration (Tejeria et al., 2002), Alzheimer's (Mendez et al., 1992; Roudier et al., 1998; Cronin-Coulomb et al., 2000), Huntington's (Janati, 1985), and Parkinson's (Dewick et al., 1991; Cousins et al., 2000) disease. As a disorder, the term should be limited to cases with selective deficits in face recognition, when this is disproportionately severe compared to other visual or cognitive dysfunction.

These subjects have both a problem discriminating known from unknown faces and a bias to experiencing most faces as unfamiliar. To cope with their recognition difficulty they identify people by voices or nonfacial visual cues, such as gait or mannerisms. They can use distinct facial cues such as unusual glasses, hairstyle, or scars, which circumvent the need to recognize the whole face.



Fig. 9.2. Apperceptive general visual agnosia. Magnetic resonance axial images from a 62-year-old woman with posterior cortical atrophy, showing reduction in occipital gyri. She presented with one year of difficulty reading, searching for items, identifying money and faces, and reading the numbers on the phone dial.

As with healthy subjects, the context of an encounter can aid recognition, so that they recognize a colleague at work but not on the street (Young and Ellis, 1989; Kracke, 1994; Takahashi et al., 1995). Some subjects have an anterograde form, in that they recognize old acquaintances but not people met after the onset of their lesion (Tranel and Damasio, 1985; Young et al., 1995). Most prosopagnosic subjects are aware of their problem and its social difficulties, except for some patients with childhood onset (Young and Ellis, 1989; de Haan and Campbell, 1991; Kracke, 1994). Some subjects find their disability distressing and are severely dysphoric.

Confirmation of impaired face recognition usually involves a battery of photographs of public persons, as in the famous faces test (Albert et al., 1979). Ideally such a test should include unfamiliar faces to determine whether the subject can discriminate known from unknown faces (Barton et al., 2001). Also ideally, failure should be contrasted with intact recognition through other means, such as famous voices or famous names. Interpretation of the results of famous-faces tests must take into account the cultural and social background of the subject. Tests of short-term recognition for recently seen faces can circumvent the question of prior long-term exposure to famous people. These include the faces subtest of the Warrington recognition memory test (Warrington, 1984) and the Cambridge face memory test (Duchaine and Nakayama, 2005), which require subjects to indicate which of a set of faces were ones they had seen in a previous phase of the test.

Identity is not the only type of information we derive from faces: we also extract data about factors such as gaze direction, emotional expression, age, ethnicity, and sex. Some prosopagnosic subjects are impaired in processing these other aspects also (Young and Ellis, 1989; Campbell et al., 1990; de Haan and Campbell, 1991; Kracke, 1994; Stephan et al., 2006; Humphreys et al., 2007), but in others the face-processing defect appears to be fairly specific for identity (Bruyer et al., 1983; Tranel et al., 1988; Sergent and Villemure, 1989; Sergent and Poncet, 1990; Evans et al., 1995). Evidence from functional imaging and monkey studies suggests that areas that encode facial identity and facial social signals may be separate (Perrett et al., 1992; Gauthier and Logothetis, 2000; Haxby et al., 2001), providing support for hypotheses that these functions can be dissociated in prosopagnosia, provided if the lesion is selective enough for regions processing identity (e.g., the fusiform face area) and spares those processing expression (e.g., superior temporal sulcus and amygdala).

Despite the professed inability of prosopagnosic subjects to recognize faces, in some there remains some unconscious or covert face recognition (Bruyer, 1991; Young, 1994; Barton, 2009). Covert face familiarity or knowledge has been shown with physiological measures such as electrodermal skin conductance (Bauer, 1984; Tranel and Damasio, 1985; Bauer and Verfaellie, 1988) and visual-evoked potentials (Renault et al., 1989). Behavioral methods have also been used, such as forced-choice guessing of which face belongs to a name (McNeil and Warrington, 1991; Sergent and Signoret, 1992; Barton et al., 2001), the speed to learn to pair names with famous versus anonymous faces (Bruyer et al., 1983; McNeil and Warrington, 1991; Sergent and Signoret, 1992; Schweinberger et al., 1995), scanning eye movements when viewing famous faces (Rizzo et al., 1987), and priming and interference effects from faces upon tasks that involve classifying names (de Haan et al., 1987b; Young et al., 1988). Current hypotheses about covert recognition suggest that it represents either the residual function of a damaged face-processing network (Farah et al., 1993; O'Reilly and Farah, 1999; Young and Burton, 1999; Barton and Cherkasova, 2003), or, particularly in the case of residual autonomic indices, a parallel pathway involving the superior temporal sulcus and amygdala (Tranel et al., 1995).

Prosopagnosia frequently forms a quartet with three other clinical findings: a visual field defect, achromatopsia or hemiachromatopsia in those with fusiform lesions, and topographagnosia. The field defect is commonly a left or bilateral upper quadrantanopia but sometimes a left homonymous hemianopia (Levine et al., 1985; Rizzo et al., 1987; Takahashi et al., 1995; Barton et al., 2004). These four findings are not present in every subject but are a loosely associated quartet, reflecting the extent of damage among neighboring structures in the medial occipital lobe. Some prosopagnosic subjects also have a mild visual object agnosia (Young and Ellis, 1989; de Haan and Campbell, 1991). Those with more anterior temporal damage can have visual or verbal memory disturbances (Landis et al., 1986; Bauer and Verfaellie, 1988). Other occasional deficits include simultanagnosia (Bruyer et al., 1983; de Haan et al., 1987a), palinopsia, visual hallucinations, constructional difficulties, and left hemineglect (Landis et al., 1986; Takahashi et al., 1995).

There are two main issues in the pathophysiology of prosopagnosia. The first is whether the disorder arises from damage to a module or network that is dedicated to the processing of faces alone (Kanwisher, 2000), or whether it reflects damage to an expertise network required for making subtle differentiations between similar exemplars of the same object category, of which faces are merely the most dramatic and universal example (Tarr and Gauthier, 2000). Thus a key question is whether prosopagnosic subjects show deficits in making such within-category distinctions for other object classes. The early reports have been mixed. Some subjects cannot identify types of car, food, or coin, or specific exemplars of buildings, handwriting, or personal clothing (Lhermitte et al., 1972; Whiteley and Warrington, 1977; Damasio et al., 1982; de Haan and Campbell, 1991), while others can identify personal belongings (de Renzi, 1986), individual animals (Bruyer et al., 1983; McNeil and Warrington, 1993), specific places (Bruyer et al., 1983; Evans et al., 1995), cars (Bruyer et al., 1983; Henke et al., 1998), flowers (Evans

et al., 1995), vegetables (Henke et al., 1998; Riddoch et al., 2008), and eyeglasses (Farah et al., 1995).

Face-specificity in prosopagnosia is a complex issue, however. On the one hand, given the size and variability in natural human lesions, one can always argue that any problem with recognizing other objects may be due to damage to adjacent structures rather than to the processes required for face recognition. On the other hand, claims that other object recognition is spared can be countered by assertions that testing was not detailed enough. For example, measures of reaction time in two prosopagnosic subjects showed deficits in nonface processing even when accuracy rates were normal (Gauthier et al., 1999). Also, one must somehow find a level of differentiation and individuation for other objects that is appropriate to the prosopagnosic subject's premorbid expertise: better bird recognition should be expected of bird fanciers than of those who have no interest in these creatures, for instance. Few objects have as universal an interest to humans as faces. Nevertheless, one study of vegetable and fruit recognition found impairments in almost all prosopagnosic subjects (Barton et al., 2004; Barton, 2008b).

The second issue concerns the staging of the functional deficit in prosopagnosia. In cognitive models, face recognition is decomposed into a series of stages (Bruce and Young, 1986): visual processing generates a face percept, which is then matched to a memory store of previously encountered faces. A successful match activates person-identity nodes with biographical and semantic data about people, nodes which can also be accessed through other nonfacial routes, such as voice or gait processing. Prosopagnosia can theoretically arise from dysfunction at any stage, although it is probable that, in most subjects, especially those with large lesions, damage may not be limited entirely to one stage. Nevertheless, classical accounts divide prosopagnosia into two broad classes: (1) failure to form a sufficiently accurate facial percept (apperceptive prosopagnosia); and (2) inability to match an accurate percept to facial memories (associative prosopagnosia) (Damasio et al., 1990; de Renzi et al., 1991).

In apperceptive prosopagnosia, the subject cannot form an accurate picture of faces. Previously this diagnosis was inferred from nonfacial tests, such as overlapping figures, silhouettes, gestalt completion tests, and global texture patterns (Levine and Calvanio, 1989; Rentschler et al., 1994; Evans et al., 1995; Takahashi et al., 1995). However, it is not clear that these probe skills relevant to face recognition. Recent studies using facial stimuli suggest that apperceptive prosopagnosia is marked by defects in processing facial structure, including the precise spatial arrangement of the features within a face (Barton et al., 2002; Joubert et al., 2003) and holistic forms of face processing (Bukach et al., 2006). In some subjects there is a regional face-specificity, with particularly poor discrimination in the eye region (Caldara et al., 2005; Bukach et al., 2006, 2008; Barton, 2008b), which is normally the most informative facial area for extracting identity.

In associative prosopagnosia, there is failure of perceptual data to access to face memory stores (Tranel and Damasio, 1985; Damasio et al., 1990; de Renzi et al., 1991). In some cases this may be because of a disconnection between facial percepts and the memory stores (Takahashi et al., 1995; Fox et al., 2008a). In others the facial memories may be lost. The diagnosis of associative prosopagnosia has traditionally been indirect, based on demonstration of intact face perception. A more direct probe of the status of facial memories is imagery (Takahashi et al., 1995; Barton and Cherkasova, 2003). Loss of imagery for famous faces is particularly prominent in prosopagnosic subjects with anterior temporal lesions (Barton, 2008b).

Just as prosopagnosic subjects can have a variety of functional deficits, they can have a variety of lesions on imaging. There are three main categories of prosopagnosic lesion. The classical case is bilateral damage to the lingual and fusiform gyri of the medial occipitotemporal cortex (Meadows, 1974b; Damasio et al., 1982) (Fig. 9.3). Functional magnetic resonance imaging (fMRI) studies show that faces activate several regions, including the superior temporal sulcus, an occipital face area, and the fusiform face area (Kanwisher et al., 1997; McCarthy et al., 1997; Haxby et al., 2000) (Fig. 9.4). It is likely that the occipital face area and the fusiform face area play critical roles in processing facial identity. Analysis of lesions in apperceptive prosopagnosia suggests common involvement of the right fusiform gyrus (Barton et al., 2002; Barton, 2008b); fMRI of one well-studied subject showed that lesions had destroyed the right occipital face area and the left fusiform face area (Rossion et al., 2003; Schiltz et al., 2006). A second lesion category is a unilateral right occipitotemporal lesion (de Renzi, 1986; Landis et al., 1986; Michel et al., 1986; Sergent and Villemure, 1989; Schweinberger et al., 1995; Takahashi et al., 1995), most of which also involves the right fusiform gyrus (Barton et al., 2002). A third type of prosopagnosia is associated with right or bilateral anterior temporal lesions (Evans et al., 1995; Barton and Cherkasova, 2003; Barton et al., 2003b). Rarely, there are cases with unilateral left occipitotemporal lesions, most often in subjects who are left-handed, who may therefore have anomalous lateralization of face processing (Mattson et al., 2000; Barton, 2008a).

The relation between structural lesions and functional deficits in prosopagnosic subtypes has been studied. It is hypothesized that anterior temporal lesions cause the



Fig. 9.3. Apperceptive prosopagnosia. Magnetic resonance coronal images of a 41-year-old man showing bilateral inferior occipital lesions from head trauma 20 years previously, affecting the region of the right fusiform face area (arrow). He has prosopagnosia, dyschromatopsia, and right hemianopia.

associative form of prosopagnosia, whereas occipitotemporal lesions affecting the fusiform and/or occipital face areas may cause the apperceptive form (Damasio et al., 1990; Barton et al., 2002; Barton and Cherkasova, 2003). The difference between unilateral and bilateral occipitotemporal lesions is a subject of research.

The most common causes of prosopagnosia are posterior cerebral artery infarctions, head trauma, and viral encephalitis (Damasio et al., 1982; Takahashi et al., 1995; Barton et al., 2002), partly because of the potential of these lesions to cause bilateral damage. Tumors, hematomas, abscesses, and surgical resections are less frequent, but common among patients with unilateral lesions (Malone et al., 1982; Landis et al., 1986; de Renzi et al., 1991). Progressive forms occur with focal temporal atrophy (Tyrell et al., 1990; Evans et al., 1995; Joubert et al., 2003). Prosopagnosia can be a transient manifestation of migraine (Martins and Cunha e Sa, 1999). There is increasing interest in a developmental form of prosopagnosia also (McConachie, 1976; Young and Ellis, 1989; de Haan and Campbell, 1991; Kracke, 1994; Ariel and Sadeh, 1996; Barton et al., 2003a; Duchaine and Nakayama, 2006), which may be associated with social developmental disorders such as Asperger syndrome (Kracke, 1994).

There is no known treatment for prosopagnosia. One subject reportedly learned new faces when asked to rate faces for a personality trait or remember semantic data about them, but benefit did not transfer to other views of the same faces (Polster and Rapcsak, 1996). An attempt to enhance expert processing by training a prosopagnosic subject to recognize artificial objects of high similarity (Greebles) improved their recognition of such objects but worsened their face recognition (Avidan et al., 2005). Hence, this trained expertise did not transfer to face processing but competed with it. Otherwise, subjects may benefit from learning adaptive skills, such as using nonfacial and nonvisual cues more effectively in identifying people.

Other disorders of face perception

The last element in the cognitive model of face processing is the person-identity node, containing biographical information about individuals. Because biographical data can be accessed from several routes, this leads not to prosopagnosia, in which subjects can still recognize people from other sensory cues, but to a people-specific amnesia, in which no cues can prompt recollection of other people, while other types of memory remain intact. This has been described with right temporal pole lesions (Ellis et al., 1989; Hanley et al., 1989; Evans et al., 1995; Gainotti et al., 2008). This localization is consistent with functional imaging studies showing that name and face recognition both activate the anterior middle temporal gyrus and temporal pole (Leveroni et al., 2000; Gorno-Tempini and Price, 2001).

Older reports have shown that right hemispheric lesions can impair some aspects of face processing in subjects who are not overtly prosopagnosic. There are reports of defective perceptual matching of unfamiliar faces (de Renzi et al., 1968; Carlesimo and Caltagirone, 1995), though a study that examined both accuracy and reaction times for famous and nonfamous faces in subjects found that the perceptions of familiar and anonymous faces were not truly independent processes (Young et al., 1993b). A less-studied aspect of face processing is the analysis of dynamic facial information, such as gaze, expression, and age. The monkey and



Fig. 9.4. The core face-processing network. Functional magnetic resonance images in a healthy subject showing axial (top), coronal (middle), and sagittal (bottom) images. The red regions indicate significantly greater blood oxygen level-dependent signal when viewing faces than when viewing objects. From left to right, in the right hemisphere, are the occipital face area, the fusiform face area, and the superior temporal sulcus. (Courtesy of Chris J Fox.)

functional imaging data would suggest that these are more likely to be associated with damage to the superior temporal sulcus (Campbell et al., 1990; Haxby et al., 2000). While an older study suggested that a selective defect for facial expression occurred with left hemispheric lesions (Young et al., 1993b), a more recent study using fMRI confirmed that damage to the right superior temporal sulcus can affect the processing of facial expression selectively (Fox et al., 2008b).

Some subjects mistake strangers for people known to them (Young et al., 1993a; Rapcsak et al., 1994, 2001). False recognition of faces has been described mainly with large middle cerebral artery strokes, affecting lateral frontal, temporal, and parietal cortex (Young et al., 1993a; Rapcsak et al., 1994). Some of these subjects are impaired in discriminating familiar from unfamiliar faces, yet deny problems in recognizing people (Rapcsak et al., 1994). Others have intact face recognition (Young et al., 1993a; Rapcsak et al., 1996). Most have right prefrontal damage, which may impair self-monitoring and decision-making, leading to premature judgments of facial similarity from fragmentary data, with failure to reject incorrect matches (Rapcsak et al., 1996).

DISORDERS OF READING

Acquired alexia is loss of reading ability in a literate person. Reading requires low-level visual processes such as good foveal resolution, complex visual functions such as pattern and form perception, accurate visuospatial and ocular motor skills to shift of attention and fixations during line scanning, and competent linguistic analysis. Not surprisingly, many lesions and disturbances can impair reading.

To assess an alexic subject, first one measures visual acuity with simple stimuli such as the directional Es of the Snellen chart or high-contrast gratings, to avoid a confound with impaired letter processing. Other elements of aphasia must be assessed, including naming and writing. Reading aloud and for comprehension can be tested informally with any available material: premorbid intellect and reading proficiency must be taken into consideration. Analysis of errors can be useful: taking more time with longer words indicates a letter-by-letter strategy in spelling dyslexia; mistakes limited to the left side of text or words point to neglect dyslexia; and semantic substitutions or phonological errors are characteristic of central dyslexias (Black and Behrmann, 1994). Standardized assessments exist. The Wide Range Achievement Test has subjects read aloud words of increasing difficulty until they make a string of errors. To test comprehension, the Chapman–Cook speed of reading test has subjects read paragraphs and cross out the word that is incongruous in meaning.

Pure alexia (alexia without agraphia)

Subjects with pure alexia can write but cannot read well, despite good acuity and oral and auditory language skills. At the severe end of the spectrum, subjects with global alexia (Binder and Mohr, 1992) cannot read numbers, letters, or other abstract symbols, such as musical notation for pitch, road signs, and map symbols (Horikoshi et al., 1997; Beversdorf and Heilman, 1998). At the mild end, letter-by-letter readers can read but slowly and with occasional errors, perhaps evident only by comparison with controls of similar educational level (Black and Behrmann, 1994). The characteristic sign of this spelling dyslexia is the word length effect, in that the time needed to read a word increases with the number of its letters (Bub et al., 1989; Coslett et al., 1993). Reasons for the word length effect continue to be debated. Some argue that it reflects a switch from a normal rapid parallel processing of all letters in a word to a slow serial process (Rayner and Johnson, 2005). Others suggest that it reflects impaired perception of letters, requiring time and effort to distinguish letters that are highly similar to other letters (Fiset et al., 2005).

There is evidence of covert processing in pure alexia. Some subjects can rapidly indicate whether a string of letters forms a word or not (Albert et al., 1973; Coslett and Saffran, 1989; Coslett et al., 1993), point to words they cannot read aloud (Caplan and Hedley-White, 1974), or identify letters more quickly when they are embedded in real words than in random letter strings (Bub et al., 1989). Some can categorize words semantically (Coslett et al., 1993) or match words to objects (Albert et al., 1973; Feinberg et al., 1995). Their eye movements show effects of word frequency and predictability, indicating some partial access to "top-down" linguistic data (Johnson and Rayner, 2007).

Pure alexia is frequently associated with a right hemianopia or superior quadrantanopia, sometimes with hemiachromatopsia (Damasio and Damasio, 1983; Lepore, 1998). Subjects often have color anomia (Geschwind and Fusillo, 1966; Damasio and Damasio, 1983) and sometimes anomia for other objects. Anomia affects items heard or felt as well as seen, indicating its linguistic rather than visual origin (de Renzi et al., 1987). There may be impaired verbal memory, other visual agnosias, or a disconnection type of optic ataxia, in which the right hand has difficulty reaching for objects in the left visual field (Damasio and Demasio, 1983; de Renzi et al., 1987).

Almost all lesions causing pure alexia are in the left hemisphere. Most are located in the medial and inferior occipitotemporal region (Damasio and Damasio, 1983; Binder and Mohr, 1992). The most frequent cause is left posterior cerebral artery stroke, but other causes include primary and metastatic tumors (Greenblatt, 1973; Vincent et al., 1977; Uitti et al., 1984), arteriovenous malformations (Ajax, 1967; Bub and Arguin, 1995), hemorrhages (Henderson et al., 1985), herpes simplex encephalitis (Erdem and Kansu, 1995), cysticercosis (Verma et al., 2004), multiple sclerosis (Jonsdóttir et al., 1998; Mao-Draayer and Panitch, 2004), Creutzfeldt–Jakob disease (Adair et al., 2007), and posterior cortical atrophy (Freedman et al., 1991; Beversdorf and Heilman, 1998).

Two major explanations exist for pure alexia. Some cases may represent a disconnection of the visual input in both hemifields from language areas in the left hemisphere (Dejerine, 1892; Geschwind and Fusillo, 1966). Most commonly, a left occipital lesion produces a complete right hemianopia and extends anteriorly to the splenium, forceps major, or white matter around the occipital horn (Damasio and Damasio, 1983; Lanzinger et al., 1999), interrupting callosal fibers from the intact right occipital lobe. Visuolinguistic disconnection may result from the combined effects of bilateral occipital lesions in subjects with bilateral field defects, without splenial damage (Lepore, 1998). Less commonly, lesions of the white matter underlying the left angular gyrus may disconnect visual input to language processors, even without right hemianopia (Greenblatt, 1973; Vincent et al., 1977; Erdem and Kansu, 1995). Support for disconnection derives from unusual cases in which pure alexia results from the combination of a splenial lesion and a right hemianopia from nonoccipital lesions, such as left geniculate infarction (Silver et al., 1988; Stommel et al., 1991) or demyelination of the left optic radiations (Mao-Draayer and Panitch, 2004).

Other cases of pure alexia may represent a selective visual agnosia from damage to the left fusiform gyrus (Damasio and Damasio, 1983; Kleinschmidt and Cohen, 2006; Leff et al., 2006). Functional neuroimaging shows that this region contains an area that responds selectively to letters or words, the 'visual word form area' (McCandliss et al., 2003) (Fig. 9.5). Subject studies with fMRI and diffusion tensor imaging have shown



Fig. 9.5. The visual word form area. Coronal and axial image from a healthy English-speaking subject showing regions with greater activity when viewing English words than Korean script that is not familiar to the subject. Arrow indicates the visual word form area on the coronal image, which is also the area activated on the axial image. (Reproduced from Barton et al. (2010), by permission of MIT Press.)

that disconnection or destruction of the visual word form area can impair reading (Molko et al., 2002; Gaillard et al., 2006; Martin, 2006; Epelbaum et al., 2008). Stimulating this region through subdural electrodes can induce pure alexia in epileptic subjects (Mani et al., 2008). However, lesions in alexia do not always affect the visual word form area but sometimes involve other regions such as the middle temporal gyrus (Price and Devlin, 2003; Reinke et al., 2008).

Additional support for a visual agnosia in at least some cases comes from pathological reports inconsistent with disconnection. One subject had a lesion of the left fusiform and lingual gyrus but no splenial degeneration (Beversdorf et al., 1997). Another subject without hemianopia had a lesion in the left lateral occipitotemporal cortex, too ventral to interrupt fibers to the angular gyrus (Benito-León et al., 1997).

Functionally, apperceptive defects (Rentschler et al., 1994; Behrmann et al., 1998), simultanagnosic-like defects (Shallice and Warrington, 1977; Levine and Calvanio, 1978; Warrington and Shallice, 1980; Mendez and Cherrier, 1998), and associative defects (Vaina et al., 1994; Chanoine et al., 1998) have all been proposed as the basis for a word form agnosia. The word length effect, the letter-by-letter strategy, and greater difficulties with handwritten script and briefly shown words suggest an inability to grasp words as a whole (Warrington and Shallice, 1980). Studies of letter-byletter reading show that these subjects can only encode letters in words serially (Rayner and Johnson, 2005). Further evidence that perceptual knowledge of word forms is impaired in some alexic subjects with medial occipital lesions comes from studies showing that their supposedly preserved writing ability also displays problems with words with irregular spelling, which cannot be written from their sound alone using spelling rules, and require access to an internal dictionary (Rapcsak and Beeson, 2004; Sheldon et al., 2008).

As with faces, there are two main theories about the basis of a word form agnosia. One is that it represents a domain-specific disorder, affecting only words (Warrington and Shallice, 1980; Cohen and Dehaene, 2004). The other is it is the most obvious manifestation of a more general visual impairment (Farah and Wallace, 1991; Behrmann et al., 1998). Pure alexia can be associated with impaired processing of local textural features (Rentschler et al., 1994) and impaired identification of complex objects in line drawings (Behrmann et al., 1998). Recent studies show that alexic subjects have slower recognition and reduced perceptual span for both letters and digits, arguing against a letterspecific defect (Ingles and Eskes, 2008; Starrfelt et al., 2009).

The prognosis for alexia is variable and depends on the underyling pathology. Global alexia can resolve into spelling dyslexia (Lanzinger et al., 1999). Improvement may depend upon increased perceptual tuning for letters in the right fusiform region, to compensate for the damaged left visual word form area (Henry et al., 2005). One fMRI study showed that improvement was associated with increased activation in regions around the visual word form area and in the superior parietal lobule, suggesting reorganization of a wordprocessing network (Ino et al., 2008).

Many innovative rehabilitative strategies are being developed, though none yet proven (Leff and Behrmann, 2008). These include altering text to highlight the spacing between words or phrases (Beeson and Insalaco, 1998; Maher et al., 1998), enhancing oral articulation during reading (Conway et al., 1998), repetitive oral reading of text (Beeson and Insalaco, 1998), attempts to enhance implicit or covert processing of whole words (Maher et al., 1998; Sage et al., 2005), and finger tracing of letters in subjects presumed to have a disconnection syndrome (Maher et al., 1998; Nitzberg Lott and Friedman, 1999). Successful strategies may need to be tailored to the specific defect of a given subject.

Hemialexias

The disconnection hypothesis for pure alexia (Dejerine, 1892; Geschwind, 1965) requires two disconnections of the left angular gyrus, one for each hemifield. Each has also been described in isolation. In left hemialexia, reading is impaired in the left hemifield only, because of isolated damage to the splenium or the callosal fibers elsewhere (Gazzaniga and Freedman, 1973; Molko et al., 2002). This disconnection has been visualized in one subject with a combination of fMRI and diffusion tensor imaging (Molko et al., 2002). Right hemialexia has been reported with a lesion of the left medial and ventral occipital lobe (Castro-Caldas and Salgado, 1984). Left hemiparalexia is a rare syndrome reported with splenial damage after surgery for arteriovenous malformations (Binder et al., 1992). Subjects make substitution and omission errors for the first letter of words, much like neglect dyslexia (see below), but they do not have hemineglect, and have left-sided lesions with right hemianopia rather than the converse.

Alexia with agraphia

In this disorder both reading and writing are impaired but oral and auditory language is preserved. Alexia with agraphia is associated with lesions of the left angular gyrus (Dejerine, 1892; Benson, 1985) (Fig. 9.6) or sometimes the adjacent temporoparietal junction (Kawahata and Nagata, 1988; Paquier et al., 2006). It is more likely a linguistic than perceptual disorder: its characteristics suggest a deep dyslexia with deep dysgraphia (see below) (Glosser and Friedman, 1990; Cohen et al., 2000; Sheldon et al., 2008). It may be accompanied by other elements of Gerstmann's syndrome and some degree of anomia (Paquier et al., 2006). It has been described with tumors (Sheldon et al., 2008), posterior cortical atrophy (Ardila et al., 1997), and Marchiafava–Bignami disease (Ferracci et al., 1999).

Older reports noted that subjects with Broca's aphasia from left frontal lesions have trouble with all expressive language output, and therefore also with reading aloud and writing. This is not surprising, but some also have marked difficulty understanding written material, despite relatively preserved comprehension of spoken language (Benson et al., 1971; Benson, 1977). Such subjects are better at occasionally grasping a whole word, while unable to name its

Fig. 9.6. Alexia with agraphia. Computed tomographic axial images of a 55-year-old man with a glioblastoma in the left hemisphere, affecting the angular gyrus, among others. He had acalculia, a mild anomia, a right hemianopia, and a deep dyslexia with agraphia. (Reproduced from Sheldon et al. (2008), by permission of the *Canadian Journal of Neurological Science*.)

constituent letters, hence the name "literal alexia" or "letter blindness." Careful study does show that these subjects have impaired comprehension of syntax in written or spoken language, similar to the agrammatism of their verbal output; hence their alexia and agraphia form part of a broader linguistic deficit.

Secondary alexia

Visual loss and reading. Bilateral reductions of central acuity of any cause impair reading ability; this will not be missed on proper examination. Visual field



defects that do not affect central acuity can impair reading too, though. Bitemporal hemianopia can cause hemifield slide, in which the absence of overlapping regions of binocular visual field leads to unstable binocular alignment with transient duplication or disappearance of words during reading (Kirkham, 1972). Homonymous field defects cause hemianopic dyslexia (Schuett et al., 2008) when the central 5° is affected (Trauzettel-Klosinski and Brendler, 1998; Trauzettel-Klosinski and Reinhard, 1998). Overall reading speed is more prolonged for subjects with right hemianopia than for those with left hemianopia (de Luca et al., 1996; Trauzettel-Klosinski and Brendler, 1998). With languages written from left to right, subjects with left hemianopia have trouble finding the beginning of lines, since the left margin disappears into the field defect as they scan rightwards (Zihl, 1995; Trauzettel-Klosinski and Brendler, 1998). Marking their place with an Lshaped ruler helps. Right hemianopia prolongs reading times, with more fixations and smaller saccadic amplitudes as they read (Zihl, 1995; de Luca et al., 1996; Trauzettel-Klosinski and Brendler, 1998). As in pure alexia, hemianopic subjects take longer to read words with more letters, but this word length effect is milder. They read single words better than sentences, unlike pure alexic subjects, and imaging shows that the left fusiform gyrus is involved in pure alexia but not in hemianopic alexia (Leff et al., 2006). Smaller type and learning to read obliquely with the page turned nearly 90° may help. Reading performance can improve with time as both right and left hemianopic subjects learn adaptive strategies (Trauzettel-Klosinski and Brendler, 1998). A well-designed study has shown that practice with an optokinetic approach using text scrolling to the left improves reading speed in alexia with right hemianopia (Spitzyna et al., 2007).

Attention and reading. Subjects with left hemineglect from right parietal or frontal lesions make left-sided reading errors, known as neglect dyslexia (Behrmann et al., 1990). They omit the left side of lines or pages, and with individual words make left-sided omissions ("bright" read as "right"), additions ("right" read as "bright), or substitutions ("right" read as "light"). Vertically printed text is not affected (Behrmann et al., 1990). The impairment represents a combination of both a space-centered deficit, in which text on the left side of space is ignored, and an object-centered deficit, in which the left sides of words are ignored. Rarely, it may occur without other signs of hemineglect (Patterson and Wilson, 1990).

An attentional dyslexia has been described in which the perception of single items is adequate, but perception of several objects simultaneously is impaired (Shallice and Warrington, 1977; Levine and Calvanio, 1978). These subjects identify single words normally but not several words together, and identify single letters but cannot name the letters in a written word. When reading they make literal migration errors, in which a letter from one word is substituted into another word ("poor baby" read as "boor baby"). Letters are mistaken for others that look similar ("o" and "c") (Levine and Calvanio, 1978). This dyslexia has been reported with lesions in the left parietal lobe (Shallice and Warrington, 1977) or temporo-occipital junction (Levine and Calvanio, 1978). This entity bears resemblance to some modern cases of visual word form agnosia associated with fusiform damage.

Eye movements and reading. Abnormal fixation and saccades may impair reading. The acquired ocular motor apraxia from bilateral frontal or parietal lesions can impair reading severely (Holmes, 1918b; Pierrot-Deseilligny et al., 1986; Husain and Stein, 1988; Baylis et al., 1994). Brainstem or subcortical lesions may cause profound saccadic and fixation abnormalities: reading difficulty with progressive supranuclear palsy has been linked to square-wave jerks disrupting fixation and hypometric and slow saccades impairing scanning (Friedman et al., 1992a). Their downgaze paresis also makes reading hard.

Central dyslexia

These deficits stem from impaired linguistic processes. Central dyslexias are formulated in terms of parallel processing channels in reading models derived from cognitive neuropsychology (Black and Behrmann, 1994). After letters or words are identified visually, there are at least two means of processing. One is a direct phonological route, in which generic pronunciation rules convert a string of letters into sound. The other is an indirect lexical route, in which the whole word is perceived and identified in an internal dictionary of written words, which then generates its pronunciation. Subjects with phonological dyslexia have lost the direct route and cannot deduce the pronunciation of pseudowords or words they have not seen before (Beauvois and Dérouesné, 1979; Funnell, 1983; Friedman and Kohn, 1990; Friedman, 1995). Subjects with surface dyslexia have lost the indirect route and cannot pronounce irregular words like "yacht" and "colonel" (Shallice et al., 1983; Patterson and Morton, 1985; Cummings et al., 1986; Friedman et al., 1992b). Deep dyslexia resembles phonological dyslexia, but subjects substitute words with a similar meaning for the correct one ("boat" read as "ship") (Coltheart, 1980).

Topographagnosia

These subjects get lost in familiar surroundings. With a complex task like route-finding, this symptom may have a number of different causes, many of which require further elucidation (Aguirre and D'Esposito, 1999). One form associated with prosopagnosia and achromatopsia (Malone et al., 1982; Bauer, 1984; Landis et al., 1986; Sergent and Villemure, 1989; Young and Ellis, 1989; de Haan and Campbell, 1991; Evans et al., 1995) is an inability to identify familiar landmarks and buildings, 'landmark' agnosia (Takahashi and Kawamura, 2002). This occurs with right ventral temporo-occipital lesions (McCarthy et al., 1996; Pai, 1997). The origins of landmark agnosia are debated. Some propose that it reflects a selective multimodal memory disturbance rather than a strictly visual problem (McCarthy et al., 1996). However, functional imaging shows that buildings and places activate a specific region in occipitotemporal cortex, the parahippocampal place area, which is adjacent to the fusiform face area (O'Craven and Kanwisher, 2000). Lesions here could create an agnosia for landmarks, and explain the frequent association with prosopagnosia.

Recent work also shows that the hippocampus and retrosplenial cortex are involved when subjects form and use a mental map of their environment (Iaria et al., 2007). Cognitive map formation is one of the most efficient and flexible ways to orient within the world. Congenital topographagnosia can be associated with impaired cognitive map formation and failure to activate the hippocampal locus seen in healthy subjects (Iaria et al., 2009).

The spatial processing needed to describe, follow, or memorize routes can be disrupted by right parietotemporal lesions (de Renzi et al., 1977; Pai, 1997). The key deficit may be an egocentric disorientation, in which subjects cannot represent the location of objects and buildings with respect to themselves (Aguirre and D'Esposito, 1999).

Another form is a heading disorientation, with failure to represent direction with respect to cues in the external environment, rather than in relation to the subject. This has been associated with posterior cingulate lesions (Takahashi et al., 1997). This may have also been the case in a subject with a left parahippocampal and retrosplenial lesion who had defective route-finding associated with alexia and other severe visual amnestic deficits (Sato et al., 1998).

Parahippocampal lesions have also been implicated in an anterograde topographagnosia, in which new routes cannot be learned, though old routes are still known (Habib and Sirigu, 1987).

DORSAL OCCIPITOPARIETAL PATHWAY DISORDERS

Cerebral akinetopsia

Cerebral akinetopsia is a selective impairment in motion perception. Only two cases of akinetopsia from bilateral lesions have been well described, LM and AF. LM has been the subject of many reports (Zihl et al., 1983, 1991; Hess et al., 1989; McLeod et al., 1989; Baker et al., 1991; Shipp et al., 1994; Rizzo et al., 1995; Campbell et al., 1997; Marcar et al., 1997). Symptomatically, LM had no impression of motion in depth or of rapid motion (Zihl et al., 1983). Fast targets appeared to jump rather than move (Zihl et al., 1991). Subjects with motion deficits from unilateral lesions are either asymptomatic or have more subtle complaints, such as "feeling disturbed by visually cluttered moving scenes" and trouble judging the speed and direction of cars (Vaina and Cowey, 1996; Vaina et al., 1998).

Tests for motion perception require computeranimated displays that are not available in most clinics. It is not possible to infer perceptual deficits solely from impaired motor responses to moving stimuli, such as pursuit eye movements. Although LM and AF had impaired smooth pursuit (Zihl et al., 1983), subjects with unilateral lesions impairing motion perception may have normal smooth pursuit, and conversely subjects with abnormal pursuit may have normal motion perception (Barton et al., 1996a).

Many different aspects of motion perception can be tested. Even with extensive bilateral lesions, not all motion perception is lost. Distinguishing moving from stationary stimuli is still possible (Zihl et al., 1983) and the contrast sensitivity for moving striped patterns is almost normal (Hess et al., 1989). LM could discriminate the direction of small spots (Zihl et al., 1991) and random dot patterns in which all dots were moving in the same direction (Baker et al., 1991; Rizzo et al., 1995). However, LM and AF had trouble perceiving differences in speed, and their perception of direction was severely affected when even small amounts of random motion or stationary noise were added (Baker et al., 1991; Vaina, 1994; Rizzo et al., 1995).

These deficits are reflected in a number of perceptual tasks involving motion cues. When searching among multiple objects for a target, LM could not restrict her attention to moving objects (McLeod et al., 1989). LM and AF could not identify two-dimensional shapes defined by differences in motion between the object and its background. LM was also impaired for threedimensional shapes defined by motion (Vaina, 1994; Rizzo et al., 1995). When lip-reading, LM had trouble with polysyllables uttered rapidly, and her judgment of sound was biased by auditory rather than visual cues (Campbell et al., 1997). On the other hand, LM could easily see biological motion (e.g., identifying the movements of a human body).

LM suffered sagittal sinus thrombosis with bilateral cerebral infarction of lateral occipitotemporal cortex (Zihl et al., 1991). AF had acute hypertensive hemorrhage with similar bilateral lateral occipitotemporal lesions (Vaina, 1994). In monkeys, motion-specific responses are found in areas V5 (middle temporal area) and V5a (medial superior temporal area), in the superior temporal sulcal region (Zeki, 1991). The lateral occipitotemporal area has been identified from histological markers and functional imaging as homologous to monkey area V5 (Clarke and Miklossy, 1990; Watson et al., 1993; Tootell and Taylor, 1995; Barton et al., 1996b). The correspondence of this region to monkey V5 is strengthened by a study showing similar patterns of deficit and spared abilities in LM and monkeys with V5 ablations (Marcar et al., 1997). Stimulation of the putative location of V5 in humans causes impairments in motion perception (Blanke et al., 2002; Cowey et al., 2006).

Unilateral lesions of the human V5 area (Fig. 9.7) cause more subtle abnormalities of motion perception. Some small series report contralateral hemifield defects for speed discrimination (Plant et al., 1993; Greenlee et al., 1995), detecting boundaries between regions with different motion, and discriminating direction amidst



Fig. 9.7. Hemiakinetopsia. Magnetic resonance axial image from a 23-year-old man 1 year after he suffered a hemorrhagic infarction of the right lateral occipitotemporal lobe (small arrow). He had impaired ipsilateral pursuit and abnormal motion perception. (Reproduced from Barton et al. (1996a), by permission of Oxford University Press.)

motion noise (Barton et al., 1995). As in LM and AF, motion detection and contrast thresholds for motion direction are normal (Plant et al., 1993; Greenlee et al., 1995). At present, there are few data on hemispheric differences. While an earlier study found a predominance of right-sided lesions (Vaina, 1989), similar defects have been identified subsequently with damage to either side (Regan et al., 1992; Barton et al., 1995).

Are different types of motion perception affected by different lesions? Studies show that subjects can differ considerably in the pattern of preserved versus affected motion processes (Vaina et al., 2005). Firstorder motion refers to stimuli in which motion can be computed by correlating the spatial distribution of luminance in the visual scene over time. However, we can discern motion from other information besides luminance, such as contrast, texture, stereopsis, and flicker. These are known as second-order motion. Initial case studies suggested that first- and second-order motion may have separate loci, with second-order motion affected by a lesion near the V5 region (Vaina and Cowey, 1996; Vaina et al., 1999) and first-order motion by a medial occipital lesion affecting V2 and V3 (Vaina et al., 1998), but recent studies have found that deficits of first- and second-order motion perception co-localize to the V5 region (Greenlee and Smith, 1997; Braun et al., 1998). Some segregation of firstand second-order processing is still possible, though, as impaired first-order and preserved second-order motion perception were occasionally seen in subjects with smaller peri-V5 lesions (Greenlee and Smith, 1997). An fMRI study (Smith et al., 1998) suggested that signals from second-order motion first emerge in V3 and VP, and may be later integrated with first-order motion signals in V5.

Other studies contrasting individual cases have suggested that lesions in the V5 region may selectively impair the integration of motion signals over larger areas, as suggested before (Barton et al., 1995), while medial lesions affecting the V3 region may impair judgment of speed or the detection of boundaries between regions of different movement (motion segregation) (Vaina et al., 2005). Another potential distinction is between these short-range processes and the more long-range integration of position data that gives rise to apparent or "high-level" motion perception. Defects in high-level motion perception arise from parietal rather than occipitotemporal lesions, and may be related more to defects in transient visual attention than the processing of motion signals (Batelli et al., 2001, 2003). Likewise, the perception of biological motion, as when one derives the percept of a walking motion from point sources of light attached to a body, is impaired primarily by lesions not of V5 but of the more

anterior superior temporal polysensory area (Vaina and Gross, 2004).

The relation of pursuit eye movements to motion perception is of interest. During pursuit, the fMRI signal related to motion perception is enhanced in V5 and in a more dorsal parieto-occipital location (Barton et al., 1996b; Freitag et al., 1998). Some of the neural activity in area V5a during pursuit may be information about the eye movement itself (efference copy). Since movement of images on the retina can be generated either by moving objects while the eye remains still, or by eye movement while the world remains still, efference copy may serve to disambiguate the two. A subject with vertigo induced by moving objects could not take his own eye movements into account when estimating object motion (Haarmeier et al., 1997). He had bilateral occipitoparietal lesions, possibly of a homolog of V5a.

Other defects can be associated with akinetopsia. The proximity of the optic radiations means that hemianopic defects are frequent. The two akinetopsic subjects had large lesions and, not surprisingly, defects on other perceptual tasks. AF was poor at recognizing objects seen from unusual angles and in incomplete line drawings, and on spatial tests such as hyperacuity, line orientation, line bisection, spatial location, and stereopsis. LM had poor perception of forms constructed from cues of texture, stereopsis, or density (Rizzo et al., 1995).

The prognosis of motion perceptual deficits is still unclear. Two cases showed significant improvement over 6–12 months (Barton and Sharpe, 1998; Braun et al., 1998). In monkeys the pace and degree of recovery are correlated with the size of the lesion and the extent of damage to both V5 and V5a (Yamasaki and Wurtz, 1991). Recovery with larger lesions presumably reflects adaptation involving other surviving motionresponsive regions of cortex.

Bálint syndrome

Bálint's syndrome (Bálint, 1909; Hecaen and de Ajuriaguerra, 1954) is a loosely associated triad of visuospatial dysfunctions: simultanagnosia, optic ataxia, and ocular motor apraxia.

Subjects have a deficit in attention, in that they cannot attend to more than one or at most a few objects at a time. Thus they have trouble with visual search tasks (Coslett and Saffran, 1991) maintaining attention over large regions of space (Rizzo and Robin, 1990), and cannot count the number of objects present (Demeyere et al., 2008). Even with a single object they may have trouble reporting on more than one of its attributes (Coslett and Lie, 2008b). This attentional defect may also underly simultanagnosia, the inability to interpret a complex scene with multiple interrelated elements, despite intact perception of the individual elements (Wolpert, 1924). What constitutes an element or object is a complex matter, depending not only on visual properties but also on cognitive factors. For example, such subjects can identify single letters but have difficulty identifying multiple letters in a random string. However, if that string is a word or even a pronounceable nonword, performance is better, indicating that the letters have been grouped into a single linguistic element (Baylis et al., 1994). Similarly, improvement in detecting multiple objects improves if they are semantically related (Coslett and Saffran, 1991; Coslett and Lie, 2008a). Related to simultanagnosia is the phenomenon of local capture, in which subjects fail to see the global layout despite seeing individual local elements, as with hierarchical Navon letters, which are large letter patterns made from the arrangement of small letters (Huberle and Karnath, 2006; Shalev et al., 2007) or shapes defined by patterns of moving dots (Huberle et al., 2009). With stimuli that have a compelling global interpretation, such as faces, they can show the opposite, global capture (Dalrymple et al., 2007).

An additional spatial problem is visual disorientation (Holmes, 1918b), a defect in judging the spatial position and distance of objects. This may contribute to optic ataxia, which is difficulty in reaching to visual targets despite normal limb strength (Luria et al., 1962) and position sense. Misreaching may represent more than just visuospatial misperception, since it can affect one arm more than the other (Bálint, 1909) and affect reaching to somatosensory targets, such as the subject's own body parts (Holmes, 1918a; Blangero et al., 2007). Laboratory measures of reaching, pointing, and grasping have shown increased latency, abnormal hand trajectories, increased variability of the end of the reach, tendency to reach to one side, as well as dissociations of distance and direction control (Perenin and Vighetto, 1988; Jakobson et al., 1991; Rizzo et al., 1992). In optic ataxia, reaching is usually normal towards foveated objects and impaired for objects in peripheral vision, and shows a correlation of reaching errors with target eccentricity (Bonner-Jackson et al., 2005; Himmelbach et al., 2006). Also, reaching is characteristically worse when performed immediately to the target: a delay of a few seconds can improve both reaching accuracy (Danckert and Rossetti, 2005; Himmelbach and Karnath, 2005) and the ability to avoid obstacles during movement (Rice et al., 2008). This suggests that the parietal cortex may play a specific role in rapid visuomotor control, and that there are alternative routes using the ventral stream for calculating object location to guide slow reaching movements. This is supported by observations that subjects with visual agnosia have normal rapid reaching responses but are impaired if action is delayed for a few seconds (Goodale et al., 1994). Additional studies in subjects with unilateral optic ataxia show that the reaching errors occur in a gaze- or eye-centered map, rather than a body frame of reference (Khan et al., 2005, 2007; Dijkerman et al., 2006).

The ocular motor abnormalities of Bálint's syndrome are not well characterized. There are probably several components. Psychic paralysis of gaze or acquired ocular motor apraxia indicates a difficulty in initiating voluntary saccades to visual targets (Hecaen and de Ajuriaguerra, 1954; Cogan, 1965). While reflexive saccades to suddenly appearing visual objects or noises are normal, these subjects may have difficulty making a saccade on command. A related problem is spasm of fixation (Holmes, 1930). Today this is defined as a problem with initiating saccades away from a fixation point that remains visible (Johnston et al., 1992; Nyffeler et al., 2005). Once saccades are generated, there may be gross inaccuracies in saccadic targeting, causing the eyes to make a series of wandering saccades in search of the target, which is still visible (Holmes, 1918b; Luria et al., 1962). Visual exploration of more complex scenes also shows gross disorganization of searching fixation patterns (Nyffeler et al., 2005).

In addition to these core features, there can be other abnormalities in spatiotemporal processing. The spatial mapping difficulties that underlie inaccuracies in reaching or saccades can also be accompanied by failures to integrate spatial information from different spatial maps in a multimodal fashion, as seen in experiments that present simultaneous visual and tactile stimuli (Valenza et al., 2004). Impaired judgment of the temporal order of stimuli has been named "sequence agnosia" (Malcolm and Barton, 2007). Subjects can show migration of features of one object to another, indicating coarsening of the encoding of feature location (McCrea et al., 2006).

To diagnose Bálint's syndrome one must exclude more general cognitive dysfunction, hemineglect, and extensive visual field defects. Perimetry can be difficult, due to inattention, fatigue, and erratic fixation (Mackworth, 1948; Broadbent, 1958; Mackworth et al., 1964). Many reports of Bálint's syndrome are marred by inadequate documentation of visual fields: extensive peripheral scotomata leaving only "keyhole vision" (Luria, 1959) can create signs that mimic all components of the Bálint triad. Simultanagnosia is usually tested by asking the subject to report all items and describe the events depicted in a complex visual display with a balance of information in all quadrants, such as the cookie theft picture from the Boston Diagnostic Aphasia Examination (Goodglass and Kaplan, 1983). Subjects will omit elements and fail to grasp the story being shown. At the bedside, subjects can be asked to pick up a number of coins scattered on a table (Holmes, 1918b). To test for optic ataxia, easily seen items are placed at different locations within arm's reach of the subject, who is asked to touch or grasp them, with each hand tested separately for each side of hemispace (Castaigne et al., 1975). With unilateral lesions, the problem tends to be worse for reaching with the contralateral hand or in contralateral hemispace. Misreaching for visual targets is contrasted with reaching to parts of the subject's own body, though more diffuse disturbances in parietal spatial representation may impair both (Holmes, 1918a). Such generalized misreaching can be confused with cerebellar dysmetria, but the latter is usually accompanied by intention tremor and dysdiadocokinesia. Ocular motor apraxia is confirmed by comparing the subject's difficulty in making saccades to command with their ease in making reflexive saccades to sudden targets in the natural environment, such an unexpected noise.

The relationship between the different elements of Bálint's syndrome has been debated. In the past simultanagnosia has been held responsible for both optic ataxia and ocular motor apraxia (Luria et al., 1962). Recent reports show that deficits in peripheral attention occur with optic ataxia (Striemer et al., 2007), and that impaired visuomotor control of fast movements is common to both saccades and reaching in optic ataxia (Gaveau et al., 2008). However, others consider that each element of the triad is potentially dissociable (Holmes, 1918b; Hecaen and de Ajuriaguerra, 1954; Luria et al., 1962; Cummings et al., 1986). Even when there are simultaneous deficits in attention and optic ataxia, their spatial characteristics may diverge, suggesting a degree of independence (Striemer et al., 2009). Therefore the ocular, reaching, and attentional disturbance may each have a different pathophysiological origin, even if they can also contribute to each other's manifestations. The incapacity to combine elements into a whole seen in simultanagnosia was thought to reflect an inhibitory action of a focus of attention upon surrounding regions (Luria, 1959). Failure of long-range spatiotemporal processes to sustain and distribute attention is likely (Rizzo and Robin, 1990; Coslett and Saffran, 1991). Reaching under visual guidance may be mediated by recursive processes involving a cerebral sensorimotor network (Battaglia Mayer et al., 1998), which can be disrupted at a number of key points, including parietal and frontal cortex (Boller et al., 1975; Nagaratnam et al., 1998). Inaccurate saccades and difficulty initiating saccades reflect damage to specific structures involved in saccadic control, such as the lateral intraparietal area (Andersen et al., 1992) and/or the frontal eye field.

There are often other disturbances. These include a variety of bilateral hemifield defects, usually affecting the lower quadrants more severely, and other visuospatial defects such as left hemineglect, akinetopsia, and astereopsis (Holmes and Horrax, 1919; Hecaen and de Ajuriaguerra, 1954; Rizzo, 1993). Smooth pursuit is often impaired. Subjects may complain of distorted perception, with metamorphopsia, micropsia, and macropsia, or visual perseverations such as palinopsia and monocular polyopia. Visual agnosias may be present with more extensive lesions (Kertesz, 1979). Visual-evoked potentials may be normal (Onofrj et al., 1995) or abnormal (Kertesz, 1979; Jarry et al., 1999), probably depending upon the extent of associated damage.

Bálint's syndrome results from bilateral occipitoparietal damage (Fig. 9.8). The early reports emphasized the role of the angular gyri, though the lesions clearly were more extensive, involving the splenium, white matter, and pulvinar (Bálint, 1909; Holmes, 1918b). Modern imaging links simultanagnosia to lesions of the dorsal occipital lobes in Brodmann areas 18 and 19 (Rizzo and Robin, 1990). The lesions of optic ataxia are more variably localized, including premotor cortex, occipitoparietal regions, cortex inferomedial to the angular gyri (Rizzo et al., 1992), and occipital-frontal white-matter connections (Auerbach and Alexander, 1981; Nagaratnam et al., 1998). These can be unilateral (Perenin and Vighetto, 1988; Ando and Moritake, 1990; Nagaratnam et al., 1998). In the occipitoparietal lobes, the classical location is considered the intraparietal sulcus and superior parietal lobule (Perenin and Vighetto, 1988), though a meta-analysis has suggested a more inferior location, at the junction between the inferior parietal lobule and the superior occipital cortex (Karnath and Perenin, 2005). Acquired ocular motor apraxia in its dramatic form requires bilateral lesions of the frontal eye fields, inferior parietal lobes, or both (Holmes, 1918b; Luria et al., 1962; Pierrot-Deseilligny et al., 1986), though it has been described in one subject after unilateral pulvinar resection (Ogren et al., 1984).

The most common causes of Bálint's syndrome are ischemia, particularly from watershed infarctions (Montero et al., 1982; Hijdra and Meerwaldt, 1984) or vasculitis (Jacobs et al., 2004; Malcolm and Barton, 2007), and degenerative disorders such as Alzheimer's disease (Hof et al., 1990; Graff-Radford et al., 1993), subacute sclerosing panencephalitis (Yapici, 2006), and posterior cortical atrophy (Perez et al., 1996; Iizuka et al., 1997; Beversdorf and Heilman, 1998; Jarry et al., 1999). Other causes include tumors, abscesses, trauma (Kertesz, 1979), leukoencephalopathies, Marchiafava–Bignami disease (Trobe and Bauer, 1986; Truffert et al., 1996), prion disorders, and, in subjects with acquired immunodeficiency syndrome (AIDS), progressive multifocal leukoencephalopathy (Paytubi Gari et al., 1998) and human immunodeficiency virus (HIV) encephalitis (Schnider et al., 1991). Recurrent transient episodes can occur rarely with migraine (Shah and Nafee, 1999).

The prognosis varies with etiology. Subjects with acute infarction can recover significantly with time, whereas those with posterior cortical atrophy deteriorate. Recovery can be dissociated, with ocular motor deficits improving but not attentional abnormalities (Nyffeler et al., 2005). Little is known about treatment. Cognitive and perceptual rehabilitative approaches involving verbal cues and organizational search strategies have been reported to improve visual function and reaching in three subjects (Perez et al., 1996).

Astereopsis

One of the important clues to distance from the observer is the disparity between the retinal images of the object in the two eyes. Astereopsis occurs in subjects with bilateral occipitoparietal lesions (Holmes and Horrax, 1919; Rizzo and Damasio, 1985). Milder deficits occur with unilateral lesions, and there may be other associated visuospatial defects. Subjects may complain that the world looks flat and that they cannot tell the depth of objects, and they may misreach for objects in depth but not in direction. Whether astereopsis can explain all these symptoms can be debated, as there are many other monocular clues to distance besides stereopsis, including relative differences in object size and intensity (which artists exploit), and differences in object motion as the observer's head moves sideways, along the interaural axis (motion parallax). Whether these other depth perceptual functions are also impaired in these subjects needs further investigation. Stereotests, which are cards viewed with different polarized or colored glasses worn by the two eyes, can diagnose deficient stereopsis (Patterson and Fox, 1984).

BLINDSIGHT

Some remaining visual processing can be demonstrated in at least some subjects who have impaired conscious awareness of visual stimuli. There are two overlapping aspects of this phenomenon. One is the emphasis on the dissociation between awareness and function. In blindsight, sometimes termed "type I blindsight" (Weiskrantz, 1998), this is complete: subjects deny any awareness of a visual event (Sanders et al., 1974; Weiskrantz, 1987; Stoerig and Cowey, 1997). Other subjects retain some awareness, and hence have residual vision (Barbur et al., 1980, 1993; Blythe et al., 1987), sometimes termed "type II blindsight." The distinction between this and a severe relative hemifield deficit is not clear. The distinction



Fig. 9.8. Bálint's syndrome. Magnetic resonance axial images of a 48-year-old woman with bilateral occipital and parietal infarcts from primary cerebral vasculitis, causing a left inferior quadrantic defect, simultanagnosia, optic ataxia, and ocular motor apraxia.

between blindsight (type I) and residual vision (type II) is also not necessarily sharp, as awareness can vary along a spectrum from absence to complete certainty, encompassing various degrees of vague sensations of something being there (Marcel, 1998; Overgaard et al., 2008). Stimulus parameters can be manipulated to generate residual vision in some trials and blindsight in other trials in the very same subject (Weiskrantz et al., 1995; Sahraie et al., 1997).

The second aspect is anatomical, in that blindsight studies are often considered the investigation of remnant visual function following loss or deafferentation of striate cortex. This relates to debates about the substrate of consciousness. In both monkey and human studies, some investigators have concluded that conscious visual perception requires striate cortex (Celesia et al., 1991; Merigan et al., 1993; Cowey and Stoerig, 1997), though this is contradicted by claims that hemispherectomized subjects can retain awareness in the blind hemifield (Ptito et al., 1991).

Experimentally, blindsight studies can use either direct techniques, which require a response to the blind stimulus, which can be detection, forced-choice discrimination, localization, or some other response, or indirect techniques, in which the experimenter determines whether responses to seen stimuli are influenced by stimuli in the blind field. Regardless of method, a taxonomy of blindsight has been proposed that groups blindsight phenomena into three clusters that may relate to different anatomical substrates (Danckert and Rossetti, 2005). 'Action blindsight' includes manual and saccadic localization, 'attention blindsight' covert orienting, and inhibition of return and motion perception, and 'agnosopsia' encompasses abilities to report on properties such as color, form, and semantic content.

Action blindsight

The first demonstration of blindsight found a weak correlation of saccade amplitude and target position in blind hemifields, mainly for a limited range of paracentral locations (Pöppel et al., 1973), as seen in subsequent reports (Sanders et al., 1974; Weiskrantz et al., 1974; Perenin and Jeannerod, 1975). However, in some cases localization was reported for regions that later recovered on perimetry (Weiskrantz, 1987) and several studies have failed to find saccadic localization in the absence of awareness (Meienberg et al., 1981; Blythe et al., 1986; Barton and Sharpe, 1997b). Using an indirect approach, a study of hemianopic subjects showed that saccadic trajectories to a seen target deviate with distractors present in the blind hemifield (Van der Stigchel et al., 2008). Targets in blind hemifields have been localized by manual reaching and pointing too. In some studies this is weak and variable, sometimes only present with residual vision (Perenin et al., 1980; Blythe et al., 1987; Corbetta et al., 1990), but there are other reports of nearly normal manual localization (Weiskrantz et al., 1974; Marcel, 1998). Reaction times for reaching to targets in the blind field are more rapid than on trials when no stimulus is present (Cowey et al., 2008b). Most of these studies examined targets varying only in the horizontal dimension; a study with targets varying in both vertical and horizontal position found good localization with manual but not with saccadic responses (Carey et al., 2008), suggesting that these response maps are dissociable in blindsight.

Attention blindsight

Studies show some residual perception of the speed and direction of rapid bright spots (Barbur et al., 1980; Blythe et al., 1987; Morland et al., 1999). With larger stimuli such as optokinetic gratings, some subjects can discriminate motion direction (Perenin, 1991) and a few may experience an illusion of self-motion (Heide et al., 1990). An indirect strategy showed that perception of optic flow within the intact hemifield could be enhanced by optic flow in the blind hemifield (Intriligator et al., 2002). Some of these motion abilities may be derived from perception of spatial position or flicker rather than of motion, as studies with random dot stimuli that minimize these confounds found no residual motion perception (Barton and Sharpe, 1997a; Azzopardi and Cowey, 2001). Motion information might also guide eye movements. Recovery of optokinetic responses was reported in 1 subject with cortical blindness (ter Braak et al., 1971) but not in 2 others (Perenin et al., 1980; Verhagen et al., 1997) nor in 3 hemianopic subjects (Perenin, 1991). Pursuit and saccadic responses to motion were not found in subjects with medial occipital lesions sparing the lateral human motion area, though (Barton and Sharpe, 1997b).

Spatial summation occurs when two simultaneous stimuli generate faster responses than a single stimulus; temporal summation is the decrease in reaction time when a stimulus is preceded by another that provides a temporal prompt. Evidence for summation between seeing and blind fields has been found in a minority of subjects (Marzi et al., 1986; Corbetta et al., 1990; Intriligator et al., 2002; Ward and Jackson, 2002). The opposite, a distraction effect, in which targets in the blind field slow down response times to stimuli in the seeing field, has been shown for saccades but not manual reaction times (Rafal et al.,

1990). This finding could not be replicated in another study, though (Walker et al., 2000). Inhibition of return is a normal phenomenon in which a stimulus delays the detection of a target appearing in the same location a short time later. This phenomenon was also generated in the blind hemifield of one subject (Danziger et al., 1997).

Agnosopsia

Studies of temporal and spatial contrast sensitivity have yielded mixed results, even when done on the same subject, GY (Hess and Pointer, 1989; Weiskrantz et al., 1991; Barbur et al., 1994). Recent studies on GY and other subjects with hemifield defects suggest maximal detection function at low spatial frequencies of <3.5 cycles/degree and intermediate temporal frequencies of around 10 Hz (Sahraie et al., 2003, 2008; Trevethan and Sahraie, 2003). DB purportedly can detect low spatial-frequency patches in his blind field at lower contrasts than in his seeing field (Trevethan et al., 2007a).

An early report claimed that DB could discriminate large X and O forms, perhaps through orientation perception (Weiskrantz et al., 1974; Weiskrantz, 1987). Follow-up after many years showed that he can now discriminate simple shapes such as squares and circles, and even name line drawings of animals (Trevethan et al., 2007b). In contrast, others have failed to find residual form or orientation discrimination (Perenin and Jeannerod, 1975; Blythe et al., 1987; Mestre et al., 1992; Morland et al., 1996; Perenin and Rossetti, 1996). Despite this, a few subjects could align the orientation of their grasp with that of objects in their blind field (Perenin and Rossetti, 1996; Marcel, 1998; Jackson, 1999), consistent with a dissociation between pathways for object recognition and action (Milner and Goodale, 1995).

While early studies found no evidence of chromatic perception (Weiskrantz et al., 1974; Blythe et al., 1987), later studies showed some detection of colored targets (Stoerig, 1987), and evidence of color–opponent interactions in spectral sensitivity curves (Stoerig and Cowey, 1991). One subject perceived the motion of equiluminant colored spots (Guo et al., 1998) and was aware of hue, though these responses seemed to represent an average from the entire blind hemifield (Morland and Ruddock, 1997). Pupillary responses to gratings and isoluminant colors in the blind hemifield have been shown in 1 subject (Weiskrantz et al., 1998, 1999). In another study, colored stimuli caused afterimages that evoked pupillary responses in the blind hemifield of 2 subjects, despite their failure to experience a conscious afterimage (Barbur et al., 1999).

Word perception can be influenced in blindsight. The choice of meaning of an ambiguous word (i.e., light) in

the seeing field can be influenced by a word in the blind field (i.e., dark versus heavy) (Marcel, 1998). Completion effects have been reported for form perception. Two subjects had completion effects for afterimages or illusory contours such as the Kanisza triangle (Marcel, 1998). The width of one subject's grasp as he reached for an object varied with object size (Jackson, 1999). Another study used an interference task with a stimulus flanked by distractors either different or identical to the stimulus. Reaction times to seen letters and colors were prolonged by differing flankers placed in the blind field in a subject with an occipital lesion (Danckert et al., 1998).

A direct pathway between the pulvinar and the amygdala may mediate reactions to frightening stimuli (Morris et al., 1999). A study of a cortically blind subject showed that associating a visual stimulus with a painful shock caused the development of startle reflexes to that stimulus, despite the lack of conscious perception (Hamm et al., 2003). One subject could process fearful and angry faces in his blind hemifield (de Gelder et al., 1999), and this may have correlated with activity in his amygdala (Morris et al., 2001). Another cortically blind subject could guess the emotional expressions of faces better than chance (Andino et al., 2009).

Hemidecorticate subjects

Subjects lacking a cerebral hemisphere are of interest in the debate over whether blindsight requires ipsilateral extrastriate cortex. Some have found that hemidecorticate infants will look towards their blind field when a target is presented there (Braddick et al., 1992), and adults with such lesions have some residual manual localization of blind field targets (Perenin and Jeannerod, 1978; Ptito et al., 1991). Also reported are motion and form perception (Perenin and Jeannerod, 1978; Ptito et al., 1987, 1991) and a spatial summation effect, in which subjects responded faster to a seen flash when there was another simultaneous flash in the blind hemifield (Tomaiuolo et al., 1997). However, the validity of these findings is in question. Some have failed to find blindsight outside a narrow strip along the vertical meridian, which could be explained by receptive field overlap or light scatter (Wessinger et al., 1996a, b). Other studies have also shown that the residual vision in hemispherectomized subjects can be attributed to light scatter (King et al., 1996; Stoerig et al., 1996; Faubert et al., 1999; Faubert and Diaconu, 2001).

Explanations of blindsight

Current hypotheses propose several different pathways that can bypass striate cortex to generate blindsight. These may support different types of residual visual function (Danckert and Rossetti, 2005). Action blindsight such as spatial localization may involve a subcortical pathway from retina to superior colliculus (Pöppel et al., 1973; Wessinger et al., 1996a, b), possibly with further projections to posterior parietal cortex. Evidence cited for collicular involvement includes better blindsight in the temporal than the nasal hemifield (Rafal et al., 1990; Dodds et al., 2002) and blindsight with achromatic but not blue-yellow stimuli, since S-cone inputs may not be represented in the superior colliculus (Leh et al., 2006b, 2009). Attentional blindsight such as motion perception may also involve retinotectal pathways, with additional projections via the pulvinar to extrastriate regions like V5. Perception of color and form may be mediated by direct projections from surviving lateral geniculate neurons, possibly in the intralaminar (koniocellular) layer, to extrastriate cortex (Cowey and Stoerig, 1995), particularly since the colliculus lacks color opponency.

In monkey, there is some evidence to support at least the retino-tecto-pulvinar relay to extrastriate regions, particularly those involved in motion perception. Lesions of V1 do not abolish responses in V5 (Rodman et al., 1989; Girard et al., 1992; Rosa et al., 2000) or V3A (Girard et al., 1991) unless accompanied by lesions of the superior colliculus (Rodman et al., 1989; Gross, 1991). On the other hand, one study using optical imaging found that deactivation of V1 with muscimol abolished activity in V5 (Collins et al., 2005). The existence of this relay may depend on developmental factors. An early direct projection from lateral geniculate nucleus to V5 that normally regresses with development may persist in cats with striate lesions acquired in infancy (Payne et al., 1996). A similar projection has not been found in infant monkeys, though (Sorenson and Rodman, 1999).

Physiological techniques in humans have provided some support for blindsight pathways. In normal subjects, the possibility that visual signals may arrive in V5 before V1 was suggested in some studies using evoked potentials or transcranial magnetic stimulation (Beckers and Zeki, 1995; ffytche et al., 1995) but not by others (Hotson et al., 1994). Positron emission tomography (Barbur et al., 1993), magnetoencephalography (Holliday et al., 1997), and evoked responses (ffytche et al., 1996) have shown residual activation of V5 by rapid but not slow-moving stimuli in the blind hemifields of one subject. Evoked potentials suggest that affective blindsight may be associated with early responses in the superior polysensory area in temporal cortex, followed by activation of the amygdala (Andino et al., 2009).

Human neuroimaging is also starting to make contributions. An intriguing fMRI study of GY that varied stimuli to obtain responses with and without awareness showed that residual vision was associated with activation of extrastriate visual areas and dorsolateral prefrontal cortex, and blindsight (type I) with activation in superior colliculus and medial prefrontal cortex (Sahraie et al., 1997). Diffusion tensor imaging has been used to show the existence of tracts between the pulvinar and the colliculi and between the pulvinar and visual cortex (Leh et al., 2008; Lanyon et al., 2009).

Blindsight studies require rigorous exclusion of artifacts as alternative explanations of residual visual function. First, one must ensure that the subject's fixation is accurate and stable, so that the target in the blind field does not inadvertently appear in the seeing field. Eve position monitors can help, as long as both head and eye position are controlled or detected (Balliet et al., 1985). Second, light can scatter from stimuli in the blind field into the seeing field and mimic blindsight (Campion et al., 1983; King et al., 1996; Stoerig et al., 1996; Barton and Sharpe, 1997b; Faubert et al., 1999). Flooding of the seeing field with light can minimize scatter. Other strategies include control tests with stimuli at the physiological blind spot (Weiskrantz, 1987) or control subjects with pregeniculate lesions (Pöppel et al., 1973; Perenin and Jeannerod, 1975, 1978). Those who believe that extrastriate cortex is essential for blindsight use controls with thalamic lesions (Danckert et al., 1998) or hemispherectomies (Perenin, 1991; King et al., 1996). Third, most perimetric techniques only sample a small portion of the blind field: more careful testing has shown that surviving islands of vision were responsible for residual vision in some subjects (Fendrich et al., 1992; Kasten et al., 1998; Scharli et al., 1999a, b). Some claim to exclude this hypothesis of surviving striate activation by means of fMRI (Stoerig et al., 1998). However, fMRI detects only small percentage changes in MRI signal even with strong stimulation, and its analysis uses arbitrary statistical thresholds to indicate activation; this methodology is not designed for excluding weak residual activation in cortex.

Another related issue of interpretation in blindsight is whether it simply represents some form of degraded normal vision (Weiskrantz, 2009), which might be the case with remnant striate function. Reports of blindsight in subjects with optic neuropathy would be consistent with this interpretation (Wust et al., 2002). A long-standing controversy is whether blindsight results from a "criterion shift," in that subjects replied more liberally when choosing between alternatives and more conservatively when asked if they detected something, an effect that healthy subjects display (Campion et al., 1983; Meeres and Graves, 1990; Kolb and Braun, 1995). Signal detection analysis has been used to refute this for some blindsight results (Stoerig et al., 1985; Stoerig, 1987; Azzopardi and Cowey, 1997). However, criterion shift may explain some data for blindsight motion perception (Azzopardi and Cowey, 1998). A study that correlated form perception with a graded rather than a binary response for awareness showed that, in the blind field, response accuracy was related to awareness in the same way as in the seeing field, consistent with an interpretation of degraded normal vision (Overgaard et al., 2008).

Against these observations are claims of qualitative differences between normal vision and blindsight (Weiskrantz, 2009), such as retained sensitivity to luminance but not color contrast (Kentridge et al., 2007), and poor detection of S-cone inputs (purple light). One study tested GY on an exclusion task, in which subjects were required to report which of two stimuli was not shown on a trial (Persaud and Cowey, 2008). While he performed correctly in his seeing field, GY was more likely to report the stimulus shown in his blind field. However, since healthy subjects do the same with briefly presented stimuli of which they are unaware, it is not clear that this result is inconsistent with degraded normal vision.

There has been much interest in whether blindsightlike dissociations between awareness and visual function can be demonstrated in healthy subjects. Manipulations of stimulus duration can degrade awareness and generate blindsight performance in normal subjects (Meeres and Graves, 1990; Kolb and Braun, 1995; Morgan et al., 1997), but not in all studies (Robichaud and Stelmach, 2003). Use of masking techniques can similarly reduce awareness with less effect on manual reaching (Binsted et al., 2007) or form discrimination (Lau and Passingham, 2006). Particularly relevant to the debate about the anatomy of blindsight are studies using transcranial magnetic stimulation over the occipital pole of healthy subjects to create artificial central scotomata, presumably by deactivating striate cortex (Lamme, 2006). This has been done to produce dissociations between awareness of the stimulus and residual discrimination of form and color (Boyer et al., 2005), delay of saccadic but not manual button presses to seen stimuli by distractors within the scotoma (Ro et al., 2004), discrimination of happy from sad schematic faces despite lack of awareness and inability to localize them (Jolij and Lamme, 2005), and online correction of reaching trajectories (Christensen et al., 2008). As a model of striate lesions, however, transcranial magnetic stimulation has limits. For one, deactivation may not result in as complete a cessation of function as seen with a destructive lesion. For another, the results are obtained with stimulation about 100 ms after stimulus presentation, whereas the first feedforward sweep of information through striate cortex occurs at about 35 ms. While this has been used to argue that consciousness must emerge from recurrent processing involving feedback from higher visual areas (Lamme, 2001), the implication is that there is feedforward information passing through striate cortex in these experiments that would not be present in a subject with complete destruction of striate cortex.

Theories of blindsight must also account for its variability. Recent large series, representing 46 subjects in total, suggest that blindsight is rare (Barton and Sharpe, 1997a; Kasten et al., 1998; Scharli et al., 1999a). One important variable may be the extent of additional damage to the optic radiations and extrastriate cortex. However, correlations between blindsight abilities and lesion anatomy have proved elusive (Blythe et al., 1986; Marzi et al., 1986; Magnussen and Mathiesen, 1989; Barton and Sharpe, 1997a, b). A requirement for very focal striate damage is also difficult to distinguish from a need for partial striate damage (Intriligator et al., 2002), pointing back to a potential artefactual explanation.

Another potential anatomical variable is suggested by a tractography study that showed that, in 4 subjects with hemispherectomies, the 2 with blindsight had projections to visual association cortex from both superior colliculi, whereas those without blindsight had only ipsilateral collicular input (Leh et al., 2006a). Hence variability in pre-existing connectivity may determine whether a subject will have blindsight after a lesion.

Timing of the lesion may be important in whether blindsight is present. Blindsight may require neural plasticity. If so, age at onset, time since lesion, and possibly training may be important (Stoerig and Cowey, 1997). Infants or children may be more likely to develop blindsight or residual vision in both nonhuman primates and humans (Perenin and Jeannerod, 1978; Blythe et al., 1987; Moore et al., 1996; Payne et al., 1996). Some reports imply that up to half of children with cerebral blindness may have some residual vision for moving objects (Boyle et al., 2005). Not all studies have found that age matters, however (Ptito et al., 1987).

Training blindsight

Whether blindsight can be trained is controversial. Though some deny that training helps (Balliet et al., 1985; Blythe et al., 1987), others claim that practice can improve saccadic or manual localization (Zihl, 1980; Bridgeman and Staggs, 1982; Zihl and Werth, 1984a, b; Magnussen and Mathiesen, 1989). A study that trained 9 hemianopic subjects with a variety of forced-choice localization and form discrimination tasks found improvement after 5-6 months (Chokron et al., 2008). Training of motion perception may improve performance in the retinotopic region trained (Huxlin et al., 2009), and repeated training over 3 months with detection of gratings can improve contrast sensitivity in hemianopic fields (Sahraie et al., 2006). There are even claims that such training may expand visual fields (Zihl and von Cramon, 1979; Zihl, 1981; Chokron et al., 2008).

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