Cognitive visual dysfunction

Gordon N Dutton

The human visual system is made up of an exquisitely organised array of functional units which have been the subject of investigation by many disciplines, including those of optics, electrophysiology, neuropsychology, psychology, and in the realm of cognate vision, philosophy. The nature of image degradation, resulting from pathology of the eye or anterior visual pathways, is not difficult to imagine and comprehend. By contrast, the forms that visual disturbances taking result from cerebral damage are less amenable to analysis by inductive reasoning and tax the imagination.

Recent developments in neurological imaging have highlighted the structural correlates of a diverse range of cognitive visual disorders. The resultant increased understanding and awareness of the origin of these disorders provides the potential to use the remarkable plasticity and adaptability of intact unaffected cerebral function to attempt to circumvent and adapt to these complex visual problems.

During the first years of life, play and exploration using all the senses creates a progressively enlarging cross referenced memory store for all aspects of visual performance, ranging from visual resolution to image recognition and understanding in all its forms. An internalised cross referenced multi-sensory encyclopaedia of imagery and its significance is progressively built up in the mind and provides the substrate for recognition, imagination, and dreams.

A continuous process of matching of incoming visual information with the stored data takes place. A pattern match leads to recognition and reinforcement. A failure to match results in learning for the purpose of subsequent recognition. Repetition enhances recall, but lack of reinforcement renders subsequent recognition more difficult. It is thus not surprising that disturbance of this highly complex system of information storage, retrieval, and matching can cause subjective disorders of visual function at an intellectual level.

Children with multiple handicaps due to cerebral disorders may show evidence of a wide range of cognitive visual disorders, but these remain to be classified and understood. In adults, focal brain pathology can disrupt specific components of the cognitive system including those subserving vision. The resultant functional deficits provide insights into the nature of higher visual processing. Many such patients may present first to the ophthalmologist complaining of visual disturbance.

The aim of this paper is to review recent literature concerning the more common disorders of cognitive visual impairment as they affect patients likely to present to the ophthalmologist and, where appropriate, to allude to strategies which may assist in diagnosis and management.

Temporal processing

CEREBRAL MOTION PROCESSING

Area V5 in primates (also referred to as the middle temporal lobe or MT) is specialised for visual motion perception. The homologous area in humans is located posterolaterally and ventrally in the region of the occipitoparietal temporal junction. In primates, this area receives an input from the striate cortex (V1) but also receives inputs directly from the lateral geniculate nucleus and from the superior colliculus and the pulvinar nucleus. Recent work in humans has shown that intact motion perception within a blind hemifield due to occipital damage, may be subserved by this subsystem. By contrast, destructive lesions in this region in humans have been shown to impair motion perception. Focal magnetic stimulation of this region of the cerebral cortex can produce similar, but transient, impairment of motion processing.

Cerebral motion blindness or cerebral akinetopsia was first described in 1983, in a patient who had sustained bilateral cerebral lesions affecting the lateral temporo-occipital cortex and the underlying white matter. A number of similar reports have followed since then. This condition can be disabling and hazardous and mobility training merits consideration despite the visual acuity being apparently normal.

BLINDSIGHT

Persistent visual function in the impaired visual hemifield of the patient with homonymous hemianopia, due to occipital pathology, has been well described, and in certain cases may include a reflex perceptual facility for stimulus presence, location, orientation, direction of movement, and colour. Such perception generates reflex motor responses, despite markedly deficient conscious awareness for these perceptions. Blindsight can thus be difficult to understand and is the subject of controversy. A number of mechanisms have been proposed and have been recently reviewed.

There is some evidence that certain patients with homonymous hemianopia can learn to become aware of their blindsight function by training. The patients were trained to perform saccadic eye movements to targets presented in the ‘blind’ hemifield, when triggered to do so by an auditory signal. This training was found to increase conscious awareness of visual function in the hitherto blind area. The finding that this trained awareness included an ability to identify colour, form, and movement led the authors to speculate that residual striate cortex activity was facilitated by the training programme. The treatment was said to reduce visual disability.

Pursuit eye movements become smoother and more accurate when a finger is simultaneously used to follow a target. It has been suggested that such observations could also provide the basis for rehabilitation strategies for patients with homonymous hemianopia or cortical blindness, particularly if the supranuclear gaze control centres remain intact.

Few would argue that physiotherapy for motor disorders following stroke is ineffective. Analogous treatment strategies for such visual disorders thus merit further evaluation.

Visuospatial processing

IMPAIRMENT OF CENTRAL VISUAL FUNCTION

Bilateral infarction of the occipital lobes, however, leads to
cortical blindness with central visual sparing of variable degree. Sequential infarction of the occipital lobes is probably commoner than simultaneous infarction. Denial of blindness (Anton’s syndrome) is common, but does not occur in all cases. 17

SIMULTANAGNOSIA AND BALINT’S SYNDROME
Bilateral superior occipital lobe lesions can give rise to simultanagnosia. This is a condition for which, despite having intact visual acuity, the patient is only able to attend to one component of a picture at a time. It is as if there is a defect in the parallel processing of the multiple components of a visual image and is perhaps analogous to a severely damaged telephone exchange, in which only one call at a time can get through.

In one study of three such patients, the visual acuities and visual fields were found to be normal, but during sustained visual fixation, the object of regard appeared to become fragmented or disappeared from view. 18

Balint’s syndrome has been reviewed by Miller. 19 It is a condition in which bilateral pari-occipital infarction results in a triad of disorders.

1) Simultanagnosia in which the patient is neither able to take interest in, nor to be aware of objects lying on either side of the object of regard, despite the requisite visual acuity and intact visual fields for that area.

2) Psychic paralysis of gaze occurs in which the cognitive field of view is apparently restricted to being only able to attend to one target at a time.

3) Optic ataxia is characterised by misreaching, particularly with the right hand.

These features appear to form part of the clinical picture in some multiply handicapped children with brain damage. For children for whom simultanagnosia is suspected, simplification of educational material with elimination of background visual ‘noise’ may prove helpful. If the usual educational progression into smaller print size and more complex material results in educational problems, simplification and enlargement of visual information warrants consideration.

Visual inattention, hemispatial neglect, and impaired visual memory acquisition
Visual inattention is a well recognised condition, in which a stimulus in the affected visual hemifield is not detected when an identical stimulus is simultaneously presented in mirror image fashion in the contralateral hemifield. As our visual world is often symmetrical in appearance – for example, doorways, corridors, and the printed page, it can be envisaged that visual inattention may be responsible for symptomatic problems. This is another condition in which the concept of behavioural training for symptomatic dysfunction warrants evaluation.

Hemispatial neglect is a condition in which a patient is completely or partially unaware of the existence of one half of the visual, auditory, and tactile world, which may be accompanied by denial of the symptomatology. 20 Subcortical lesions affecting the thalamus and basal ganglia on the left side of the brain tend to cause aphasia, occasionally associated with right sided neglect. However, such pathology on the right side is more likely to cause contralateral neglect with attentional errors on both sides. 21 These observations provide evidence for right cerebral dominance with regard to spatial attention. 22 There is also evidence that the neglect syndrome is associated with impaired cognitive representation of hemispatial aspects of memory, in addition to disorders of contemporary perceptual events. 23 These symptomatic syndromes are often accompanied by concomitant overriding cortical dysfunction. 24

Intact temporal lobe function is required for acquisition of new memories. Bilateral infarction of the medial temporal lobes causes disabling impairment in day to day and minute to minute memory acquisition. The mnestic defect resulting from unilateral lesions is usually not permanent, but can take up to 6 months to recover. 25

Colour processing disorders
CENTRAL ACHROMATOPSIA
This may occur as a sequel to focal ischaemic damage of the fusiform and lingual gyri. 26 Complete and partial deficits have been reported. The patient is aware of colour desaturation in the contralateral visual hemifield. Although colour vision is impaired or lost, discrimination of light brightness tends to remain unaffected. 27 Despite the lesion occupying a region of brain beneath the calcarine fissure, the complete hemifield shows impaired colour appreciation, which leads to the supposition that this area is homologous to the area V4 in monkeys, which is known to subserve colour vision for the contralateral hemifield.

Visual agnosias
Positon emission tomography affords the opportunity to investigate specific visual functions, with respect to their anatomical locations in the visual cortex. 28 Cerebral activation during object recognition tasks takes place primarily in the left occipitotemporal cortex. Face recognition, however, also involves similar regions of the right hemisphere. 29 The imagination of visual objects activates some of the same regions of visual association cortex that are activated by direct visual stimulation. 30 It is thus tempting to speculate that the act of comparison between visual data input and visual memory, which must be required for recognition, takes place in a compartmentalised fashion in specific sectors of the visual association cortex. The fact that focal damage to these areas may lead to specific visual agnosias, lends weight to such a hypothesis.

The subject of visual agnosias has been comprehensively reviewed by Grusser and Landis. 31

The descriptions which are currently available in the literature concerning visual agnosias primarily comprise selected case reports of individuals in whom focal pathology has led to specific deficits. It is important to recognise, however, that multiple deficits are more common in clinical practice, but the complex nature of such problems means that the presence of additional underlying cognitive deficits may be masked initially by communication problems, such as those observed in the multiply handicapped. Detailed history taking and direct communication by mail can often reveal evidence suggestive of such problems. The results of neurological imaging studies can also be used to infer the possible presence of these deficits, which may in turn lead to specific inquiry for evidence of visual agnosic deficits.

Anosognosia, which is the lack of awareness on the part of the patient that he or she has an agnosia, means that it is often difficult to diagnose visual agnosias. Clinical suspicion is aroused, however, in patients who are complaining of inexplicable visual problems in the presence of apparently normal visual function. The finding of homonymous hemianopia or hemianopic visual inattention provides additional corroborative evidence for a potential visual agnosia.

The descriptions in the literature concerning visual agnosias tend to focus on patients who manifest ostensibly complete deficits. It is not uncommon to see patients whose problems are consistent with incomplete variants of the conditions described below. A large number of specific deficits of visual recognition have been described. Only the
more common agnosias seen in ophthalmic practice are discussed in this review.

VISUAL OBJECT AGNOSIA
This condition can occur as a sequel to a range of cerebral disorders, but is most likely to be seen by the ophthalmologist in patients sustaining a posterior cerebral artery occlusion with concomitant homonymous hemianopia. It can occur in two forms:
(a) Apperceptive agnosia in which the form of objects cannot be categorised, despite an intact ability to distinguish intensity, width, and direction.
(b) Associative visual agnosia in which intact form perception is demonstrated by an intact ability to draw an object which is seen, but in which a deficit in visual semantic processing results in failure of recognition.

When drawing ability is assessed in left or right brain damaged patients, distinct patterns of deficit are observed. Left hemisphere lesions tend to give rise to disturbances which are primarily of internal detail, whereas right hemisphere lesions disrupt the external configuration and overall orientation of a figure. Although such drawings are fragmented, the detail is preserved. Recovery tends to be more predictable for left hemisphere lesions.

The principal differential diagnosis for object agnosia is object aphasia in which there is a probable disconnection between the striate cortex and the speech centres of the left hemisphere. This results in visual recognition of objects, but an inability to name them.

PROSOPAGNOSIA
Prosopagnosia is a disability in recognising faces which occurs without loss of understanding of underlying facial expression. It has most commonly been reported as a sequel to traumatic damage of the right occipital lobe. In our clinical experience, it is a fairly common sequel to ischaemic brain disease in the elderly, for whom the disorder can be manifest in varying degrees.

The diagnosis is suspected in patients with a good binocular visual acuity who complain of poor vision, but who cannot recognise their friends until they speak. The clinical test advocated by Grüsser and Landis is to sit a known individual among a group of individuals of the same sex and racial background, and to mask or remove additional distinguishing features by covering clothing with a sheet and by replacing jewellery. The patient is asked to identify this individual without recourse to other clues. Complete failure of identification, or significant difficulty manifested by a prolonged identification time, can be observed.

In some patients prosopagnosia appears to be caused by a defect in structural encoding, in some the defect is one of impaired extraction of characteristic physiognomic features, whereas in others the problem is one of memory retrieval and matching.

The results of combined magnetic resonance imaging and positron emission tomography in normal individuals and in subjects with prosopagnosia suggest that the processing of faces is performed by an extensive neural network, which encompasses the ventromedial region of the right hemisphere from the temporal to the occipital pole. The right lingual and fusiform gyri appear to serve the role of extracting the unique physiognomic data which define each face. The right parahippocampal gyrus plays a part in memory retrieval involving biographical data relating to previously observed faces, while the right anterior temporal cortex is concerned with retrieval of biographical data not exclusively related to faces but which is required for a face to be recognised. Despite all these observations, cases of prosopagnosia have also been recorded in which the pathology has not involved such areas.

Prosopagnosia has only rarely been described in children. In the author’s experience, some multiply handicapped children, with evidence of left homonymous hemianopia or inattention hemianopia, have impaired facial recognition of family members and friends, despite having a good visual acuity. The first step in assisting rehabilitation in the adult is to explain the origin of the visual problem, as the illusion can be very disturbing for many patients, some of whom question their own sanity. For children, explanation for the apparently paradoxical visual behaviour is reassuring for the parents.

Attempts at rehabilitation by retraining recognition have not, to the author’s knowledge, hitherto proved successful. Advice concerning alternative recognition techniques using clothing, jewellery, and footwear can prove useful, and for children the regular wearing of such identifiers by family, friends, and teachers, appears to help the child integrate socially. Voice recognition, however, is not impaired. For adults, the social strategy of informing friends of poor vision may prove helpful.

TOPOGRAPHIC AGNOSIA
The right occipital lobe also serves the function of providing a sense of direction cross referenced to visual memory, and is thus required for route finding (on a large scale), and on a smaller scale for finding things which have been placed in specific locations. It is not uncommon for patients sustaining occlusion of the right posterior cerebral artery to be prone to losing things and becoming lost themselves, particularly in new environments.

Anosognosia for this disorder also occurs. In the author’s clinical experience, adults may deny that they regularly take an incorrect route and children with the disorder have never known anything different. In a patient with long standing topographic agnosia, the response to questions concerning how often the patient has to ask the way, can help lead to the diagnosis.

Rehabilitative strategies which we have found to be effective involve the construction of alternative memory substrates for commonly taken routes. We have seen one 6-year-old child with left homonymous hemianopia, who tended to get lost in his own home until colour coding of the doors was employed. The colours have since been removed, but the child is still able to remember which door is which by remembering the colours. When he is outside, his mother adopts the custom of talking constantly about his environment. One year later, he now has few problems navigating in familiar environments, presumably because he is using alternative memory cues.

ALEXIA WITHOUT AGRAPHIA
Occlusion of the left posterior cerebral artery with infarction of the left occipital lobe and disruption of the left ventral visual association cortex, combined with damage to the tracts to the left angular gyrus (which interrupts the input from the right occipital area), causes alexia without agraphia. The classic description of this disorder was in a patient with lesions of the left occipital cortex and the splenium. Combined lesions of the left lateral geniculate body and the splenium of the corpus callosum, and isolated lesions of the posterior inferior temporal lobe of the left hemisphere, have also been described as having the same effect.

Letter by letter spelling can be employed to circumvent the problem, but this is time consuming and difficult. In some cases when such patients are asked to make lexical decisions or semantic assessments concerning rapidly presented words, accurate responses can be obtained. In one such case, encouragement of the patient to make rapid semantic decisions (a function of the intact right brain) was thought to have enhanced rehabilitation.
Conclusion
Systematic and detailed investigation of patients with cognitive visual dysfunction has provided important information concerning the structure and function of the visual brain. Such detailed investigation and classification are time-consuming and impracticable in routine clinical practice for a variety of reasons. A pragmatic clinical diagnostic approach based primarily on careful history taking, supported by appropriate neurological imaging, is required if these disorders are to be identified and patients assisted in understanding and coming to terms with their visual problems. Moreover, attempts at devising rehabilitation strategies are, of course, dependent upon accurate diagnosis. Such an approach may be criticized for occasionally providing a false positive diagnosis, but it is preferable to failing to apply a diagnostic label because of a perceived requirement to carry out time consuming behavioural testing, which is, of course, necessary for scientific published reports.

GORDON N DUTTON

Tennent Institute of Ophthalmology,
Western Infirmary,
Glasgow G11 6NT
and
Royal Hospital for Sick Children,
Yorkhill, Glasgow G3 8SJ


Dutton

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