Disorders of perception

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Introduction
Broadly speaking, perception is the final stage in the visual pathway, describing the awareness that results from the recombination of information provided by parallel visual streams. This gives rise to an internal model of our environment, allowing us to navigate and categorise, attaching meaning to visual stimuli. The patient who is unable to make sense of visual information may prove to be functionally blind, despite our best efforts as optometrists to provide a clear retinal image. A bewildering array of perceptual disorders has been described in the literature, a handful of which may be encountered in primary optometric practice. Although some of the perceptual disorders described herein are relatively rare, these cases provide valuable insights into the inner workings of the normally-functioning visual system. The following article discusses some of the ways in which visual perception has been known to be affected by damage or abnormal activity beyond the primary visual cortex (V1).

Perceptual disorders with respect to site of damage
Higher visual processing (i.e. beyond V1) may be simplified into the ventral (“what”) and dorsal (“where”/“how”) visual streams (Figure 1). Damage to the ventral stream usually results in problems relating to identification or classification of visual stimuli, whereas insult to the dorsal stream typically causes difficulties relating to localisation of objects, motion perception, spatial awareness, or visually-guided action. Specific examples for each visual stream are given below.

Figure 1: Illustration showing the ventral stream (mauve area) and dorsal stream (green area) in relation to V1.

Ventral stream damage
Visual agnosia
The term agnosia literally means ‘without knowledge’. Visual agnosia describes the situation in which patients are unable to recognise stimuli visually. They may, however, be able to use other senses or logical reasoning to aid identification. Visual agnosias are classed as either apperceptive or associative. Apperceptive agnosia indicates that the patient cannot organise the physical properties of the stimulus into a structured whole. Conversely, associative agnosia describes the case in which percepts appear fully-formed and structured to the observer, but they are unable to associate the image with a
Semantic meaning from memory\textsuperscript{4,5}. Both conditions result in poor visual recognition. Visual agnosias have been documented specific to colours\textsuperscript{6}, places, faces or even categories of objects, such as living things and manmade objects\textsuperscript{7,8}.

**Prosopagnosia**

*Prosopagnosia* is a subtype of visual agnosia, specific to faces. A network of brain areas are responsible for facial recognition, comprising the occipital face area, fusiform face area, and a portion of the superior temporal sulcus\textsuperscript{9,10}. Damage to this network can cause prosopagnosia. Both associative and apperceptive variants are reported; although both types result in poor facial recognition, in associative prosopagnosia, patients are still able to discriminate between faces, but they lack the ability to identify them\textsuperscript{11}. Apperceptive prosopagnosia is presumed to affect an earlier stage in the face recognition process, as patients are neither able to identify faces, nor perform basic distinctions between them\textsuperscript{5}. A subtle hereditary form of prosopagnosia (*developmental prosopagnosia*) affects 1 in 40 people in the general population\textsuperscript{12}.

Another disorder of facial recognition is *prosopometamorphopsia*; patients report that faces appear distorted, sometimes with the distortions being limited to specific features such as the eyes. Due to the limited number of reports of this condition, the pathomechanism is not fully understood\textsuperscript{13,14}.

**Topographagnosia**

Several disorders can lead to *topographical disorientation*; a condition in which patients find themselves lost in familiar surroundings. When visual memory is the cause, the condition is known as *topographagnosia*. Visual information about landmarks, buildings and spatial layout is encoded in the parahippocampal place area, which lies medial to the fusiform face area\textsuperscript{15}. Damage to this region results in topographagnosia. Patients may benefit from strategies such as learning street names, or using a smartphone to aid navigation\textsuperscript{16}.

**Visual anomia**

Whereas visual agnosia causes patients to be unable to recognise stimuli, visual *anomia* (also referred to as *optic aphasia*)\textsuperscript{17} is a condition in which patients are incapable of *naming* objects due to damaged connections between the visual and language centres of the brain\textsuperscript{18}. Patients demonstrate the ability to recognise objects, for example, by correctly pointing to them when named by the clinician. However, this relationship only works in one direction: the patient cannot name objects pointed at by the clinician\textsuperscript{19}. As with visual agnosias, visual anomia can be specific to certain stimuli. For example, *colour anomia* is the inability to name colours. One case study detailed a patient who was able to overcome the difficulty by instead naming the football team that wore any given colour\textsuperscript{20}.

*Pure alexia* (also known as *alexia without agraphia*) describes a condition in which patients are unable to read words efficiently, despite being able to write. Patients can usually read individual letters, and in these cases, reading can be performed ‘letter-by-letter’\textsuperscript{21}. The condition may be caused by damage to the fusiform gyrus\textsuperscript{16}.

**Capgras delusion**

The Capgras delusion is a psychiatric disorder characterised by the belief that one’s friends, relatives and peers have been replaced by lookalikes\textsuperscript{22}. The condition is believed to be caused by a disconnection between the inferotemporal cortex (area IT)
and the limbic system (which processes emotion)\textsuperscript{23}. Patients recognise faces, but the lack of an emotional response causes them to believe that the individuals are imposters.

**Cerebral dyschromatopsia**

Impaired colour perception may result from brain damage in the vicinity of the putative V4 complex\textsuperscript{24}. This is known as cerebral dyschromatopsia. In some cases, the impairment may be limited to a single hemifield, in which case it is termed hemidyschromatopsia. This is a selective deficit to colour vision; form and light sensitivity are unaffected\textsuperscript{25}. In most cases, colour perception is not completely abolished; hence the term dyschromatopsia is used to distinguish from the much rarer situation of cerebral achromatopsia\textsuperscript{26}. Due to the close proximity of regions within the brain, patients with cerebral dyschromatopsia present comorbid with prosopagnosia in 72\% of cases\textsuperscript{27}.

**Dorsal stream damage**

**Akinetopsia**

Akinetopsia translates as “vision without motion”. It is an extremely rare disorder presumed to result from damage to the human homologue of area ‘middle temporal’ (hMT+)\textsuperscript{28}, causing patients to be incapable of perceiving movement of any kind. Much of our knowledge of akinetopsia comes from the study of a single individual, patient ‘LM’\textsuperscript{29}. All aspects of motion-related vision, including the performance of visually-guided actions, were impaired. She stated that “fluids appeared frozen, like a glacier, which caused great difficulty, for example, with pouring tea or coffee into a cup”\textsuperscript{28}.

**Simultanagnosia**

Simultanagnosia is an attentional disorder in which patients can only perceive a single object within a visual scene at any one time (e.g. Figure 2)\textsuperscript{30}. In some cases, patients may only be able to perceive one attribute of an object at a time, such as colour or form, but not bind them together\textsuperscript{31}. The condition is usually reported as part of Bálint’s syndrome, which results from bilateral damage to the parietal cortex\textsuperscript{32} and causes difficulties in executing voluntary saccades (ocularmotor apraxia)\textsuperscript{33} and visually-guided actions such as reaching (optic ataxia)\textsuperscript{34}. It is presumed that the regular co-occurrence of the three symptoms is due to the close proximity of separate regions in the brain, all involving spatial vision\textsuperscript{16}.
Figure 2: Artist’s impression of simultanagnosia.

**Unilateral visual neglect**
Damage to several different sites\textsuperscript{35,36} – but usually the parietal lobe – may cause a loss of awareness of one side of space. This is known as *unilateral visual neglect*. Patients may omit half of objects when drawing, or only eat food on one side of a plate. The left side is more often affected. This is because, in most right-handed individuals, the right side of space is represented by both hemispheres, whereas the left side of space is only represented by the right parietal cortex\textsuperscript{35}. The neglected hemifield may be *egocentric* (i.e. defined relative to the head/trunk), *allocentric* (concerning spatial relationships between objects) or *object-centred* (half of individual objects are neglected)\textsuperscript{37}. The condition is a loss of *awareness*, rather than a primary visual field defect, as in hemianopia.

**Astereopsis**
Depth perception may become impaired following damage to the posterior parietal lobe, even in patients with no ocular complications, amblyopia or prior history of binocular vision disorders\textsuperscript{38}.

**Hallucinations**
The term ‘hallucination’ describes any percept arising in the absence of a stimulus\textsuperscript{39}. There are two major classes of visual hallucinations: *simple* and *complex*. A *simple* hallucination consists primarily of lights, colours, lines and geometric shapes or patterns. They may appear clearly to the observer, but lack a resemblance to real-world objects. On the other hand, a *complex* hallucination is a formed percept, often resembling faces, people, objects or even entire scenes. Simple hallucinations are generally thought to arise from **bottom-up** visual processes\textsuperscript{40}, such as the waves of cortical depression thought to be responsible for migraine auras\textsuperscript{41}. Complex hallucinations are presumed to be the result of **top-down** visual influences (i.e. they are based on memory and/or prior expectations). To some extent, top-down processes mediate visual perception on a day-to-day basis, and it has been hypothesised that
hallucinations can develop (for example, in psychosis) when the balance between top-down and bottom-up influences becomes unduly biased towards top-down inference.

Hallucinations may be caused by a range of neurological conditions, such as Parkinson’s disease and dementia with Lewy bodies. Except in definite cases of Charles Bonnet Syndrome (CBS; see below), patients experiencing hallucinations should be referred for further assessment. When recording reported hallucinations, clinicians should note whether they are simple or complex, whether they are accompanied by hallucinations in any other (non-visual) sensory modality, and whether the patient is able to clearly determine the non-real nature of the percepts. Each of these clues can help to determine the underlying diagnosis.

**Migraine aura**

Although the visual auras experienced by migraineurs are often accompanied by a severe headache, nausea, photophobia and/or phonophobia, this is not always the case. A visual migraine aura occurring in isolation is called an acephalgic migraine. Migraine auras vary from individual to individual – although teichopsia (zig-zag lines akin to castle battlements; see Figure 3) are thought of as being the classic migraine aura, a much wider array of symptoms may occur, including “small bright dots”, “coloured spots of light”, hemianopia, scotomas, blurred vision and distortions such as “mosaic” fractured vision or a “heat wave” appearance.

![Figure 3: Multiple observations of a teichopsia migraine aura recorded at different time points over 30 minutes.](image)

Although very variable, in most cases a migraine aura appears in the visual periphery with a gradual onset, lasts from 5 to 30 minutes, and precedes the headache (if any) by less than 30 minutes. Migraine auras are believed to be the result of waves of neural depression (depolarisation) spreading across the visual cortex.
**Charles Bonnet Syndrome**

Optometrists will regularly encounter patients with CBS in practice. The condition is characterised by simple and/or complex visual hallucinations following significant acquired vision loss. The complexity of hallucinations is not related to the level of visual impairment. Patients will be aware that the hallucinations are not real (if a patient lacks insight into the non-real nature of the hallucinations, referral is required). The hallucinations are exclusively visual (i.e. there is no auditory component or that of any other sensory modality). Commonly reported hallucinations include photopsiae, geometric shapes, repeating grid-like patterns, distorted disembodied faces, and small people, often wearing elaborate costumes. *Palinopsia* (persistence of a previously-viewed image), *dendropsia* (branching tree-like patterns) and polyopia are also occasionally reported.

CBS has a reported prevalence varying from 11-63% in patients with low vision. The reason for this variability likely stems from the method of questioning used. Many patients are reluctant to admit to experiencing hallucinations for fear of being labelled insane. A study by Menon in 2005 used explicit, repeated questioning of 48 patients with VA worse than 6/60 and found that 63% admitted to experiencing hallucinations when directly questioned. *None* of these volunteered the symptom without first being asked. Therefore, it is crucial that optometrists always discuss CBS with any patient developing significant acquired sight loss.

Hallucinations in CBS are believed to reduce with time, but in 75% of individuals, the condition persists for five years or longer. Although the majority of patients are not adversely affected (around 7% of patients actually view them as a positive experience), a third of patients are affected negatively by the condition, either due to disruption of their daily routine, frequent fear-inducing hallucinations, or a lack of understanding about CBS and/or concerns that they are developing mental illness.

**Alice in Wonderland syndrome**

Also known as *Todd’s syndrome*, Alice in Wonderland syndrome (AiWS) is a cluster of perceptual disturbances named after Lewis Carroll’s novel, and is characterised by *metamorphopsia* (spatial distortion), *chromatopsia* (excessively saturated colour perception) and changes in the perceived size of one’s own body parts (Figure 4). Individuals with AiWS also experience *dysmetropsia*, an umbrella term given to a collection of symptoms including changes in the apparent size of objects (*micropsia/macropsia*) as well as a sense that objects are either very far away (*teleopsia*) or extremely close (*pelopsia*). As with CBS, patients with AiWS should be aware of the illusory nature of their symptoms, although the vividness of the illusion may occasionally prompt them, for example, to check their height in a mirror. Symptoms occur in transient ‘attacks’, lasting from 10 seconds to 10 minutes. Although the exact prevalence of AiWS has not been studied, it is most commonly reported in young children (average age six years), particularly around the onset of sleep. Some individuals experience AiWS as a migraine aura. In most cases, the syndrome resolves itself within weeks or months. However, AiWS can be an early sign of neurological disease; e.g. Epstein-Barr virus or brain tumour. As such, referral is necessary to rule out life-threatening causes.
Figure 4: In addition to visual symptoms, patients with Alice in Wonderland syndrome may feel as though their own body is changing shape. Illustration from Lewis Carroll’s novel by Sir John Tenniel\textsuperscript{64}.

**Anton syndrome**
Bilateral V1 damage may result in a rare condition called *Anton syndrome*. Despite being demonstrably blind, patients with this condition unequivocally deny their blindness, claiming that they can see clearly. Patients vividly describe false surroundings and are completely unresponsive to any form of visual stimulus\textsuperscript{65}. The reason for this is unknown. This is a form of *anosognosia*, meaning a lack of awareness of one’s own disability.

**Synaesthesia**
*Synaesthesia* can be simply described as a ‘mixing’ of the senses – for example, individuals may experience tactile sensations when tasting food, or see geometric shapes when listening to music\textsuperscript{66}. Up to 150 sensory ‘pairings’ have been reported. Several theories have been proposed for the existence of synaesthesia, depending on the sensory modalities involved. No consensus has yet been reached, but two popular theories include cross-activation of neighbouring areas of the brain\textsuperscript{67}, and disinhibition of feedback from brain regions involved in multisensory integration\textsuperscript{68}. Synaesthesia is
typically present from birth, but may be acquired following brain damage. The prevalence of synaesthesia is estimated to be as high as 4.4% in the general population. Most people with the condition do not realise that synaesthesia is unusual.

Cerebral visual impairment

*Cerebral visual impairment* (also known as *cortical visual impairment*, or CVI) is a non-specific term used to describe visual perceptual disturbances in individuals with diffuse brain damage. Most commonly, the term is used to describe vision in patients with cerebral palsy or hydrocephalus. In contrast to many of the conditions described above, CVI does not relate to any specific structure in the brain, nor does it imply any specific symptom. Patients with CVI may have damage to the primary visual pathways, higher perceptual function, eye movement control, or a combination of all of these. Visual performance may vary from day to day. A structured history-taking questionnaire is available to characterise the range of visual difficulties.

Summary

Much of what we know about the neurophysiology of the extrastriate cortex is derived from invaluable case studies of perceptual disorders. Many of the conditions described here are extremely rare, but provide a fascinating glimpse of the inner workings of the visual brain. Some conditions, such as synaesthesia and CBS are rather more common, but underreported, either because patients do not realise that there is anything unusual about their perception, or, they may simply be unwilling to share their experiences. It is particularly important to discuss CBS with at-risk patients, to avoid unnecessary concern. Clinicians should have a basic understanding of the range of perceptual disturbances that may occur as a result of brain damage, as perceptual disorders usually warrant onward referral.

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References


