

Visual Hallucination and Illusion Disorders: A Clinical Guide

The past century of advances in visual neurobiology and perceptual science have all but passed visual hallucinations by. Much of our assessment and management owes more to 18th century Natural Philosophy than 21st century Neuroscience. Part of the problem has been that the experiences are traditionally considered rare, of poor localising value and without associated morbidity. However, recent recognition of their true prevalence across a range of disorders¹⁻⁴ and the fact that they typically cause distress, are stigmatising, often inappropriately medicated and can precipitate institutional care, has awakened research interest. Although much has yet to be learnt, the basic and clinical science findings help formulate a structured approach to their assessment and management. What follows is a science-led guide to how one might approach a patient presenting with visual hallucinations, either occurring *de novo* without previously recognised pathology or, more typically, in the context of ongoing neurological, ophthalmological or psychiatric disease.

Defining the symptom

Rote definitions of hallucinations and illusions trip easily off the tongue, leaving the impression that the two are entirely separate entities. However, in clinical practice the difference between them is less than clear cut. Many patients who on some occasions 'see things that are not really there' will, on other occasions, 'see real things incorrectly'. Furthermore, different classes of 'incorrect seeing' (polyopia and metamorphopsic distortions, for example) seem to apply equally to hallucinated things as real things. Put another way, patients describe illusions of hallucinations - a situation which amounts to a logical impossibility. How can something not actually there, be seen incorrectly? It all amounts to a neurophilosophical nightmare from which a pragmatic clinical escape route is to pool illusions and hallucinations into a single entity - the 'positive' disorders of visual perception - treating both as one⁵. But loosening definitions with one hand requires tightening them with the other. All reports of 'seeing things' need not indicate a positive perceptual disorder. Before diving deep into the patient's visual psyche, it is worth pausing to consider whether the symptom being described is vivid visual imagery. Visual images appear in the mind's eye and are under some degree of volitional control, as opposed to hallucinations and illusions which are externally located, unpredictable and outside volition (in the sense that one cannot choose to make a hallucination of, say, a face turn into that of a chair). Termed pseudohallucinations in the European psychopathological tradition, vivid visual images carry very different aetiological implications to those of hallucinations and illusions, imagery pointing to a diagnostic spectrum which ranges from normal phenomena to clinical anxiety syndromes. Note that, confusingly, the American psychopathological tradition uses the term pseudohallucination in a different way, referring to hallucinations that are recognised for what they are (i.e. those for which the patient has insight).

Describing the phenomenology

Although traditionally ignored as a useful clinical sign, the content of a patient's visual hallucinations may help point to the underlying diagnosis and direct future management. Time spent exploring the range of perceptual pathologies experienced pays clinical dividends. While each hallucination is, in one sense, unique - no two

patients having exactly the same experience or range of experiences - take away the intricacies of detail and one is left with only two broad phenomenological syndromes, caricatured in Figure 1. In one syndrome patients will describe hallucinations from a palette which ranges from simple unformed lines, dots, colours and flashes, through more complex grid patterns and lattices to distorted faces (grotesque or gargoyle-like), unfamiliar figures in bizarre costume (often wearing elaborate hats) and extended landscape scenes. Not all the palette is experienced by every patient, some will have several types of hallucination, others just one. The experiences are typically brief, lasting seconds to minutes, silent and may be confined to a particular area within the patient's visual field. The second syndrome is derived from a different palette. Here the hallucinations range from isolated animals and figures (often familiar and without the bizarre costumes and hats of the first grouping), through extracampine hallucinations ('feeling' rather than 'seeing' an object, typically a person watching you), to multi-modality hallucinations (e.g. hearing the hallucinations talking or feeling them crawl up one's arm) and complex delusional explanations for the experiences (e.g. the neighbour's children have stolen a key to the house and wander in and out uninvited). The experiences may be brief but can last for hours or even days. Table 1 summarises key phenomenological points to help discriminate the two syndromes.

Matching syndrome to aetiology

While the details have yet to be worked out, we know why patients with visual hallucinations fall into two broad syndromes: it is because the parts of the brain involved differ between the two. Visual hallucinations and illusions relate to transient increases in activity within visual cortex, the location of the increase defining the content of the hallucination^{6,7}. Thus increased activity in face specialised cortex leads to hallucinations of faces, increased activity in colour specialised cortex, hallucinations of colour and so forth. The palette of hallucinations in a given patient thus reflects the location and extent of their susceptible cortex. The implication is that patients who experience the first syndrome have a susceptibility in different cortical regions to those with the second. Indeed, the second syndrome must extend beyond purely visual cortex to include multi-modal areas and those underlying the generation of delusions.



Dominic Ffytche is a Senior Lecturer and Consultant at the Institute of Psychiatry and has recently completed a Wellcome Clinician Scientist Fellowship. He has a special interest in visual perception and its pathologies and runs a Visual Perceptual Disorders Clinic at the Maudsley Hospital.

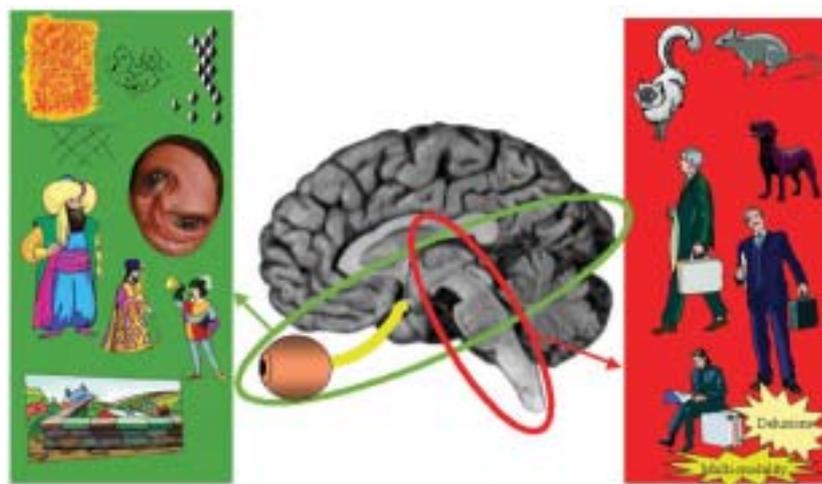


Figure 1. Visual pathway (left) and brainstem/cholinergic (right) hallucination syndromes.

The key question is what defines which cortical regions are affected in a given patient, and hence what defines which syndrome they will have. While as yet unanswered, an important clue seems to be where their primary pathology lies. Table 2 sets out the hallucination-related disorders likely to be encountered clinically, arranged into two pathophysiological groupings, one in which the primary pathology lies in the visual pathways or higher visual areas (prototypical disorder, Macular disease), the other in which it lies in the brainstem and/or cholinergic system (prototypical disorder, Parkinson's disease). The choice of terminology in the second group is not intended to diminish the possible role of other brainstem neurotransmitter systems in visual hallucinations (see⁶ for review), it serves merely to emphasise that role played by the cholinergic system⁹. Of course many hallucination-related disorders fail to fit neatly into this dichotomous classification, either because their pathology is unclear (e.g. the psychoses) or because they contain elements of both. For example, visually hallucinating patients with Lewy body dementia will have both brainstem/cholinergic pathology and Lewy bodies in higher visual areas¹⁰ while those with Parkinson's disease will have subtle visual deficits suggestive of visual pathway dysfunction¹¹. However, despite its obvious shortcomings, the classification is worth pursuing as it neatly divides clinical conditions into those associated with one or other of the syndrome palettes. Thus the spectrum of visual pathway disorders outlined in Table 1 are associated with the syndrome shown on the left of Figure 1, while the spectrum of brainstem/cholinergic disorders (as well as those of mixed and unknown aetiology) are associated with those on the right of Figure 1. One might wish to refer to the visual pathway disorder palette as the Charles Bonnet syndrome, although it is important to realise that different clinical specialties use the term in different ways⁵. The hallucinations associated with visual pathway lesions and those associated with brainstem/cholinergic lesions also differ in another important respect: their prognosis. Visual pathway hallucinations tend to be self limiting and resolve without pharmacotherapy (60% of patients with hallucinations secondary to Macular disease are hallucination free at 18 months¹²). In contrast, brainstem/cholinergic hallucinations (at least those found in Parkinson's disease) tend to persist and progress¹³, even acting to precipitate institutional care¹⁴.

Management

With such a wide range of potential aetiologies and endless possibilities for clinical overlap (glaucoma in a patient with Parkinson's disease, cognitive impairment in a patient with Macular degeneration), how should a patient with visual hallucinations be managed? Clearly the answer will depend on the clinical context; however, the general principles are summarised in Figure 2. Whatever the patient's phenomenology, it is important to review their medication to minimise anti-cholinergic load and to consider whether the hallucinations/illusions may have been precipitated by concurrent infection (often a UTI in the elderly). The question of whether to investigate depends largely on the match between their hallucinations and clinical context: if the symptoms are wrong, look for another cause. Hallucinations of a familiar dog in a patient with Parkinson's disease would not warrant further investigation but hallucinations of grid patterns confined to one hemifield might prompt imaging of the visual cortex. Similarly, prolonged hallucinations of whispering figures in a patient with Macular disease might warrant referral to an old age psychiatry service whereas a brief hallucination of an Edwardian tea party would not. Two investigations deserve special mention. Sleep studies may have a place in the investigation of brainstem/cholinergic hallucinations as these have been found to relate to REM sleep behaviour disorder and daytime REM intrusions¹⁵. Visual evoked potentials may help characterise a visual pathway lesion but there are, as yet, no known VEP markers for susceptibility to hallucinations.

Treatment options will depend on the syndrome identified. Visual pathway disorder hallucinations may be treated with reassurance and the knowledge that the hallucinations will resolve in time, although may re-occur if the visual pathway lesion progresses. The Macular Disease Society and Royal National Institute for the Blind can provide patients with further information and, in some areas, specific hallucinator support groups. Where possible, it is worth optimising visual function as this may reduce the frequency of hallucinations¹⁶. For non-distressing, visual-only brainstem/cholinergic hallucinations without persistent secondary delusions, patients may be managed with reassurance, although the experiences are likely to persist and progress. For brainstem /cholinergic syndrome hallucinations that are distressing, multi-

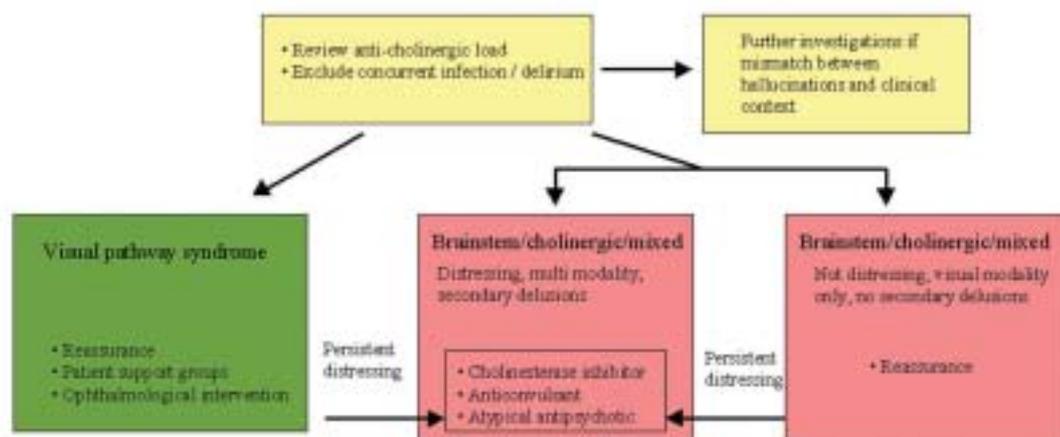


Figure 2. Management algorithm

Info on the web
Macular Disease Society
www.maculardisease.org
Tel. 0990 143 573

Royal National Institute for the
Blind
www.rnib.org.uk
Tel. 0845 766 9999

Table 1. Key phenomenological features

Content checklist	Simple forms and colours Grid and lattice patterns Disembodied faces Figures: mundane/ bizarre, familiar/unfamiliar Animals Landscapes
Duration	
Other modalities	Auditory, tactile, olfactory
Delusions/Insight	

modality or with persistent delusions (or visual pathway syndrome hallucinations that are persistent and distressing) largely anecdotal treatment options include cholinesterase inhibitors, anti-convulsants and atypical antipsychotics^{17,18}. Which class of medication to use will depend on the clinical context, with cholinesterase inhibitors a logical first choice for patients with cognitive impairment and atypical antipsychotics for those with pronounced delusions.

Future directions

While the basic science of hallucinations is beginning to inform the clinic, the converse is also true, the clinic is directing the basic science. Hallucinations and illusions that seem commonplace to the clinician pose a challenge for the basic sciences. Why should our visual systems generate spontaneous percepts of bizarrely costumed figures sporting hats? What visual function is going wrong when we see polyopic copies of the same object, neatly arranged in rows? For the astute clinician, hallucinating patients are giving us the answers to questions that have yet to be conceived.

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Table 2. Clinical associations

Visual pathway	Brainstem / Cholinergic	Mixed / unknown
Ocular pathology <i>Macular disease</i> <i>Diabetic retinopathy</i> <i>Glaucoma</i>	Iatrogenic <i>Tricyclics</i> <i>Oxybutinin</i> <i>Benztropine</i>	Dementia <i>Lewy body</i> <i>Vascular</i> <i>Alzheimer's</i>
Infarct <i>Post. cerebral artery</i>	Parkinson's disease	Psychosis <i>Schizophrenia</i> <i>Bipolar disorder</i>
Partial seizure <i>Occipital</i>	Narcolepsy	Delirium
Migraine	Peduncular lesion	Bereavement

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Correspondence to:
Dr D H Ffytche MRCP
MRCPsych
Senior Lecturer
Institute of Psychiatry PO70
De Crespigny Park
Denmark Hill
London SE5 8AF
E-mail: d.fytche@iop.kcl.ac.uk