

# Charles Bonnet syndrome: forgotten but important

Visual hallucinations occur in a number of ophthalmological, medical and neuropsychiatric disorders. Charles Bonnet syndrome is less frequently diagnosed but is a rather common cause of complex visual hallucinations in older adults. In this article, **Drs Murali Krishna** and **Peter Decalmer** review the history and discuss the assessment and management of this condition.

**H**allucination is defined as abnormal perception in the absence of an external stimulus. Visual hallucinations can occur in around 12–13 per cent of visually impaired populations<sup>1</sup> and can be elementary or complex. Complex visual hallucinations are usually well formed and often involve men, animals, and figures in bright colours and dramatic settings.

Charles Bonnet syndrome (CBS) is characterised by complex visual hallucinations generally against the background of visual impairment and in the absence of a neuropsychiatric disorder. Insight is usually preserved. Yet CBS is a less frequently diagnosed despite it being a common cause of such hallucinations<sup>2</sup>.

## History

The term CBS was introduced into medical terminology by de Morsier in 1938<sup>3,4</sup>. He named it after Charles Bonnet, the Swiss naturalist and philosopher, who in 1769 provided the first narrative description of this condition in his own grandfather. During his latter days Bonnet's grandfather, Charles Lullin, who had low vision from cataract but was otherwise psychologically sound, had experienced complex visual hallucinations of men, women, birds and buildings.

Charles Bonnet himself experienced similar phantom visions when his own vision deteriorated<sup>5,6</sup>.

## Case scenario

This scenario illustrates a clinical presentation of Charles Bonnet syndrome:

A 78-year-old gentleman is seen on a domiciliary visit at the request of the police. He is single and has never been married. A retired foundry fitter, he lives on his own in a three-bedroom semi-detached house and is well supported by his niece. He has informed the police about 'the men' he has seen walking around in his house, describing them as 'white men, six feet tall, well built and shabbily dressed'. He sees them at least twice daily. Though annoyed by their presence, he denies feeling fearful or persecuted and has made attempts to chase them away; however, he says they leave only to return. He has partial insight and does not deny the possibility of his visions being the product of his imagination.

Apart from the above mentioned vivid and graphic visual hallucinations, he has no symptoms of depression, delusions or hallucinations in other modalities. On cognitive assessment he is well oriented, without any deficits in concentration,

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registration and recall (for both short- and long-term memory). His visual impairment stems from bilateral glaucoma and age-related macular degeneration. He had been diagnosed with this condition eight years previously and is regularly reviewed by the consultant ophthalmologist. Prior to admission, his visual acuity was limited to finger count in the right eye and to 2/60 in the left. Intraocular pressures are 18mmHg in both eyes and a cupped disc ratio of 0.5 and 0.4 in the right and left eye respectively. He does not suffer from any other medical illness and his physical examination is unremarkable. He is on betaxlol eye drops and omeprazole 20mg. He is a non-smoker and there is no history of alcohol or substance misuse, nor is there a family history of mental illness. This clinical picture is suggestive of CBS and the gentleman should be admitted for a period of assessment.

### Diagnostic criteria

Universal diagnostic criteria for CBS has not

yet been established. Hence, there has been disagreement over its exact diagnostic criteria since CBS does not fit exactly into current classification systems. The nearest ICD 10 diagnosis would be organic hallucinosis. In this condition, the clinical picture is dominated by persistent or recurrent hallucinations, usually visual or auditory, occurring in clear consciousness and insight may be preserved.

The precise definition of CBS has undergone remodelling since its original introduction into medical terminology by de Morsier, who defined CBS as visual hallucinations free of optic nerve impairment. Further to his review in 1967, de Morsier himself came to a conclusion that impaired vision caused by ocular pathology was not an integral part of this condition<sup>4,7</sup>. In the absence of universal consensus, the diagnostic criteria set forth by Podoll<sup>8</sup>, and by Gold and Rabins<sup>7</sup>, are widely accepted. While Podoll's criteria provide a host of conditions to be considered and excluded to arrive at the diagnosis

**Table 1.** Diagnostic criteria**Podoll's criteria**

- > The predominant symptom is the occurrence of visual hallucinations in elderly people in normal mental health.
- > No dimming of consciousness, no dementia or debility, no organically caused affective disturbances, no delusional phenomena, no psychosis, no intoxication and no neurological disease with lesions in the central pathway of vision or the visual cortex.
- > Reduced vision (facultative but not mandatory).

**Gold and Rabins' criteria**

- > Visual hallucinations are formed, complex, persistent or repetitive and stereotyped.
- > Insight is fully or partially retained.
- > Primary and secondary delusions are absent.
- > Hallucinations in other modalities are absent.

of CBS, Gold and Rabins put emphasis on the nature of the hallucinations and the preservation of insight (*Table 1*). In addition, Gold and Rabins argue that a diagnosis of CBS does not exclude or require eye disease or brain lesions.

## Prevalence

Many people do not report having hallucinations to their doctor or optometrists because they believe they might be thought of as mentally ill<sup>6</sup>. Studies also conclude that CBS is probably underdiagnosed or misdiagnosed by both doctors and optometrists. Hence the prevalence figures reported in the literature is clearly an underestimate. Well-formed complex visual hallucinations are thought to occur in 10–30 per cent of individuals with severe visual impairment<sup>9</sup>. Prevalence of CBS in visually impaired people is thought to be between 11 per cent and 15 per cent<sup>5</sup>. It is present among one to two per cent of psychiatric outpatient attendees.

## Epidemiology

CBS is strongly associated with advanced age and bilateral visual impairment<sup>10,11</sup>. Normally it appears later on in life (mean age 76 years)<sup>12</sup>. This syndrome occurs more commonly in the elderly because of the prevalence of visual impairment in this group. CBS is equally disturbed between sexes and does not show any familial

**Table 2.** Risk factors**Risk factors for Charles Bonnet syndrome**

- > Advanced age
- > Peripheral visual impairment
- > Social isolation
- > Sensory deprivation
- > Early cognitive impairment

predisposition<sup>12</sup>. The common ophthalmological conditions associated with this syndrome are age-related macular degeneration, followed by glaucoma and cataract<sup>2</sup>. CBS can occur in people with normal vision<sup>4,8</sup>. Teunisse *et al*<sup>10</sup> found an 11 per cent occurrence of CBS in the visually impaired, but an only one per cent occurrence in those with intact vision. However, more general sensory deprivation – for example, social isolation – is an important causative factor for CBS<sup>10,11</sup>.

The characteristics of visual hallucinations in CBS are well summarised by Fernandez *et al*<sup>6</sup> and Teunisse<sup>10,11</sup>. They are exclusively visual and not associated with an auditory component or with abnormal perceptions in other modality. They are extremely vivid and well formed. They are experienced in clear consciousness. CBS patients report their visual hallucinations are always outside the body. When mobile, images tend to move en bloc. Insight is usually retained. The hallucinations do not have any clear trigger or alleviating factors. Patients have no voluntary influence on them though, in the majority, they may disappear when they close their eyes. They are unable to evoke or influence the content of the hallucinations, which can recur or remit spontaneously. Hallucinations are more common in the evening or at night.

Generally they find hallucinations a delight, pleasant or amusing. Sometimes images may stimulate anger, anxiety or even mild paranoia. They can be recurrent and persistent – subjects may experience hallucinations for a few days or for many years, and they may change in frequency and complexity.

## Mechanisms of hallucinations

The aetiology of CBS is still unknown. There are three main theories that attempt to explain

the mechanism of the hallucinations. The first and most popular is that these are 'release hallucinations' as proposed by Cogan<sup>13</sup> in 1973. He postulated that decreased visual stimulation contributed importantly to visual hallucinations in blind patients with ocular and optic nerve lesions. This theory states that reduced higher cortical centre suppression leads to a 'release' of usually inhibited 'perceptual traces'. Consequently, visual hallucinations are due to the release that result from the 'removal of normal visual afferent input to association cortex' in the visually impaired. Cogan also believes that ageing accentuates disinhibition of higher centres.

Reduced or absent visual stimulation of the visual system can lead to the increased excitability of visual cortex. Decreased vision stimulates intracerebral perceptions, as in the phantom pain syndrome. Berrios and Brook<sup>14</sup> suggested this 'deafferentation state theory' as a possible model for the visual hallucinations in CBS. The other theory is that hallucinations in visually impaired individuals occur as a part of a 'filling in' process that already exists in our brain. According to this theory, in individuals with more severely degraded vision, their brains would be acting in a manner to fill in the missing information and this results in the hallucinations they experience. This partly explains hallucinations like 'baby on the lap' or 'children in the garden'.

There may also be a contribution from the higher psychic functions including wishes, conflicts and past memories<sup>6</sup>. The possibility of epileptic activity playing a part may explain the positive results found with carbamazepine<sup>12</sup>.

## Investigations

There has been no systematic research to uncover the neuroanatomical or neuropsychological correlates of hallucinations in CBS. Most cases of CBS do not show gross or microscopic brain pathology<sup>6</sup>. Recently Plisken *et al*<sup>15</sup> studied 15 adults with CBS and found neuropsychological changes indicative of early stages of dementia as compared with age matched controls. There was no evidence of electroencephalogram (EEG) abnormalities beyond non specific slowing and no evidence of acute infarction or other lesions on magnetic resonance imaging (MRI) brain scans. However, all individuals had abnormal age corrected pattern, potentially indicative of dysfunction in the visual system.

## Differential diagnoses

Visual hallucinations occur in a number of psychiatric, medical, neurological and ophthalmologic disorders. These include:

- > delirium;
- > ophthalmological conditions – macular degeneration, cataract, glaucoma, retinal detachment and lesions of optic tract;
- > neurological conditions – Parkinson's disease, epilepsy, transient ischemic attack, cerebrovascular accident, space occupying lesions, migraine, temporal arteritis and multiple sclerosis;
- > psychiatric conditions – schizophrenia, manic depressive illness, Alzheimer's disease, multi-infarct dementia and Lewy body dementia;
- > medication – anti-Parkinson drugs, anticholinergics, corticosteroids, stimulants, benzodiazepenes, opiates and barbiturates;
- > illicit substances – amphetamines, LSD, phencyclidine (angel dust), psilocybin (magic mushroom), MDMA/ecstasy and khat chewing;
- > alcohol induced states – intoxication, withdrawal, alcoholic hallucinosis and delirium tremens;
- > bereavement;
- > rarities – peduncular, hypnogogic and hypnopompic hallucinations ('Alice in the Wonderland' syndrome).

## Management

A multidisciplinary patient-centred approach is required to manage CBS. Optimisation of vision, physician recognition, empathy, reassurance and patient education form the cornerstone of its treatment. Patients in whom the diagnosis is likely to be something other than CBS must then be referred to other specialists. Inappropriate admissions to a psychiatric ward must be avoided as it can lead to distress and stigma.

## Visual management

Hallucinations attributed to CBS may disappear after the treatment of an underlying cause of visual impairment or blindness. Some patients may respond partially – or completely – to procedures such as cataract removal. Resolution is more likely with a combination of maximising remaining vision and removing or reducing the

effects of adverse factors such as blur, glare and visual field loss<sup>6</sup>. Interestingly, hallucinations may cease as visual deterioration progresses to total blindness<sup>6</sup>. Environmental changes – for example, improved lighting – may reduce the hallucinations.

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### **Psychosocial management**

Learning that CBS is not related to mental illness often relieves patients<sup>10,11</sup>. Patients must be given an opportunity to join a psycho-education group where sufferers can be encouraged to meet, obtain reassurance and be given information and advice on specific techniques for reducing the duration of hallucinations; for example, closing or opening the eyes, blinking, putting on a light and developing distraction techniques.

Social isolation may be a contributing factor to visual hallucinations that must be addressed by promoting interpersonal contact. Attendance to day centres and involvement in friendly services may be suitable. If there is a self-help group in the locality, the patient should be encouraged to attend. A full occupational therapy assessment must be carried out and suitable measures taken to minimise risk and ensure patients live in a safe environment. In some cases, a move to sheltered accommodation or to a care home may be appropriate.

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### **Pharmacological management**

Treatment with psychotropic drugs remains controversial in CBS. Neuroleptics are only partially successful at eradicating the hallucinations. Systematic research into the pharmacological management of CBS has not been conducted. Previous studies of CBS have generally reported unsatisfactory results with medications. There have been several reports of successful treatment CBS using antidepressants (mirtazapine), typical antipsychotics (haloperidol), atypical antipsychotics (sulpiride, risperidone, olanzapine and aripiprazole), anticonvulsants (carbamazepine and valproate), cholinesterase inhibitors (donepezil), 5HT antagonists (ondanestron) and corticosteroids.

Elderly patients are particularly prone to sedation, falls, hypotension, urinary retention, extra pyramidal side effects, metabolic syndrome and cardiac side effects from psychotropics. Hence

### Key points

- > CBS is not an uncommon cause of complex visual hallucinations in the elderly.
- > Psychiatrists and ophthalmologists frequently overlook or misdiagnose this syndrome.
- > Considering the serious implications of alternative diagnoses, the correct diagnosis of this condition is imperative.
- > In addition, there is some evidence that CBS may be an early marker of dementia.

psychotropics must be used judiciously, at the lowest possible doses, with the awareness of possible side effects and drug interactions – and careful monitoring. Atypical antipsychotics have a favourable side effect profile compared to the typicals and are relatively safer in older adults.

### Prognosis and outcome

Treatment outcome with this syndrome is unclear from the literature. Prognosis varies with the nature of visual dysfunction. There are no studies that have evaluated the long-term outcome in CBS. In a series of patients, Plisken *et al*<sup>15</sup> found that some degree of cognitive deficit in CBS was very common. They proposed that isolated visual hallucinations might be an indication of early stages of dementia. Visual hallucinations as the first or the presenting symptom of dementia has been noted before<sup>17,18</sup>. Whether CBS is an early marker for dementia still needs to be established.

### Conclusion

Despite its recognition over 200 years ago, CBS has received very little attention in recent English literature. Consequently, doctors are unfamiliar with this syndrome as a possible diagnosis for complex visual hallucinations<sup>5</sup>. Ophthalmologists and psychiatrists frequently overlook or misdiagnose CBS phenomena<sup>19</sup>. The correct diagnosis of this condition is of utmost importance considering the serious implications

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of alternative diagnoses. Despite the numerous disorders associated with visual hallucinations, the most common causes in a geriatric psychiatry clinic are dementia and delirium. Clinicians assessing older patients with visual hallucinations should first carefully evaluate for these disorders<sup>1</sup>.

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