

REVIEW

## Rehabilitation in Charles Bonnet syndrome: a review of treatment options

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**Frank Eperjesi** BSc (Hons) PhD

DipOrth MCOptom FAAO

**Nabila Akbarali** BSc (Hons) MCOptom  
Neurosciences Research Institute, School  
of Life and Health Sciences, Aston  
University, UK

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People with vision loss sometimes experience visual hallucinations associated with Charles Bonnet syndrome. The appearance of these hallucinations often causes anxiety for the sufferer and can be difficult for the attending eye care professional to manage. A review of the literature highlighted a range of visual, pharmacological and social management regimes that may alleviate these hallucinations, albeit using small samples in uncontrolled trials. Eye care practitioners should be aware of methods of rehabilitation in Charles Bonnet syndrome that may lead to resolution of the visual hallucinations.

Key words: Charles Bonnet syndrome, rehabilitation, visual hallucinations, visual impairment

Charles Bonnet syndrome (CBS) is named after a Swiss philosopher who first described the phenomenon in 1769. He reported that with failing eyesight, his 89-year-old grandfather began to experience well-formed visual hallucinations of men, women, carriages and buildings, however, he was aware that they were not actually physically present. Charles Bonnet began to have a similar experience in his later life.<sup>1</sup> Although there are frequent descriptions and reports of CBS in the neurologic, psychiatric and geriatric literature, there is a sparsity of reports in ophthalmic journals, indicating limited awareness in the eye care professions.

The following diagnostic criteria are accepted by most authorities:<sup>2</sup>

1. the presence of formed, complex, persistent or repetitive, stereotyped visual hallucinations

2. full or partial retention of insight into the unreal nature of the hallucinations
3. absence of hallucinations in other sensory modalities
4. absence of primary or secondary delusions.

Other authors have claimed that impaired visual acuity and a disorder of brain function are required for CBS to develop<sup>3</sup> and there are several reports of cases in which patients experiencing CBS hallucinations have subsequently developed dementia.<sup>4</sup> Furthermore, it has been argued that insight into the unreality of the visions had to be present from the beginning or be rapidly acquired and that the hallucinations associated with CBS were better described as pseudo-hallucinations.<sup>1,5</sup> Hallucinations could precede, develop with or follow the onset of visual impairment.<sup>1</sup> Others have proposed that more general

sensory deprivation for example, due to social isolation<sup>6-9</sup> is an important contributory factor. Cases of true CBS (that is, complex visual hallucinations in the absence of a neuropsychiatric disorder and with full insight) are exceedingly rare and most cases described in the literature are CBS plus (that is, visual hallucinations in the presence of a neuropsychiatric disorder or with the sufferer totally lacking insight that the hallucinations are unreal).<sup>10</sup> A detailed review of the epidemiology, clinical characteristics and proposed pathogenesis of CBS has recently been published.<sup>11</sup>

Various prevalence rates have been reported:

1. 57 per cent of a sample of 104 with retinal or neural visual loss<sup>12</sup>
2. 10 to 30 per cent in people with 'vision deficits'<sup>13</sup>
3. 15 per cent of 319 patients with

acquired vision loss reported 'fully fledged' visual hallucinations, rising to almost 39 per cent of patients with sight loss when questioned in a context in which they felt safe<sup>14</sup>

4. eight from a group of 434 patients referred for psycho-geriatric work-up<sup>15</sup>
5. photopsias (coloured flickering lights and patterns) were experienced by 52.8 per cent of 284 consecutive low vision patients<sup>16</sup> but these are commonly reported by people with ocular pathology
6. 11 per cent in partially sighted patients<sup>17</sup>
7. 60 from 505 visually handicapped patients.<sup>18</sup>

More recently, it has been suggested that one out of seven older patients is likely to have experienced visual hallucinations.<sup>19</sup> This variability in reported prevalence rates is probably due to different definitions and diagnostic criteria as well as the reluctance of some patients to discuss their experiences for fear of being labelled emotionally disturbed.<sup>11,16,18</sup>

This review aims to describe the rehabilitation of people with CBS and, in particular, to discuss visual, pharmacological and social management. Increased awareness of this phenomenon among eye care and medical practitioners should improve its recognition and direct patients to appropriate methods of treatment.

### Visual management

Hallucinations attributed to CBS may disappear after treatment of the underlying cause of visual impairment or blindness. For example, an 82-year-old female with bilateral cataracts, who had experienced visual hallucinations, reported an improvement in VA and cessation of hallucinations following removal of the right cataract.<sup>20</sup> Similarly, hallucinations ceased for a 39-year-old male patient with proliferative diabetic retinopathy, after VA had been improved from counting fingers at 1.6 metres to 6/60 following surgical intervention.<sup>21</sup> A reduction in hallucinations has been noted after treatment of a sub-retinal haemorrhage with laser photocoagulation<sup>22</sup> and when temporal arteritis was treated with prednisolone.<sup>23</sup> Interestingly, hallucinations may cease as visual deterior-

ation progresses to total blindness.<sup>24,25</sup>

Three cases have been described in which hallucinations occurring in individuals with visual field loss, decreased VA or both, have reduced in frequency when using optical devices.<sup>26</sup> Each patient was reported to have some level of dementia and is better described as suffering from CBS plus. Optical management included trial with Fresnel prisms (later ground prism) displacing that which appeared in the left visual field to midline for a patient with left hemianopia; a 2.8x monocular telescope, combined with a patch over one eye, for a person who saw hallucinations, while watching TV; increased localised illumination during night-time reading for a patient who experienced hallucinations on waking. For another patient, with peripheral field loss due to glaucoma and central field loss due to scarred corneas and no cognitive loss, hallucinations disappeared over a sustained period of nine months using a combination of full-time spectacle wear, tinted over-shields when in bright environments and night-lights for bedroom and bathroom.<sup>27</sup>

Resolution is more likely with provision of the sharp retinal images (as with the surgical and medical interventions described above) through a combination of maximising remaining vision and removing or reducing the effects of adverse factors such as blur, glare and visual field loss. It is unlikely that resolution occurred spontaneously in these cases for two reasons. First, hallucinations returned when the interventions were halted and second, spontaneous resolution has been reported only when sight loss has progressed to blindness.<sup>24,25</sup>

### Pharmacological management

Neuroleptic agents may assist<sup>28</sup> or have no effect on CBS hallucinations.<sup>7</sup> Patients on melperone (an atypical, low-potential neuroleptic agent with minimal side effects and tolerable for older patients), experienced a reduction or total cessation of hallucinations.<sup>28</sup> In one case, the treatment was interrupted for a period of 15 days, after which the patient reported the recurrence of visual hallucinations. When pharmacological treatment was resumed,

symptoms again quickly reduced.<sup>28</sup> Haloperidol has also been reported to have limited success in treating CBS<sup>29,30</sup> or to have no effect.<sup>31-33</sup>

Other anti-seizure drugs have been reported to be successful in abolishing CBS hallucinations. One elderly patient with visual hallucinations associated with CBS for 18 months<sup>34</sup> and a 38-year-old with hallucinations but no visual impairment<sup>31</sup> were successfully treated with carbamazepine daily. It has been proposed that carbamazepine may be effective in suppressing the visual pseudo-hallucinations of CBS because of 'its inhibitory action on the increased ventral extrastriate neuronal activity in patients with CBS that persists between the attacks of hallucinatory symptoms'.<sup>35</sup> A combination of carbamazepine and clonazepam has been found to be successful,<sup>35</sup> as has 400 to 800 mg per day of valproate, which has fewer potential side effects than carbamazepine.<sup>36</sup>

One case of CBS has been successfully treated with ondansetron, a serotonin antagonist that has limited side effects and is frequently used as an anti-emetic for cancer patients receiving cytotoxic drugs.<sup>37</sup> When treatment was stopped, hallucinations promptly recurred and when treatment was resumed, the hallucinations ceased. However, research into the treatment of CBS using ondansetron and of its side effects is extremely limited. Cisapride, a drug that shares some pharmacologic properties with ondansetron, has been proposed as an alternative and two patients receiving 30 to 40 mg per day reported cessation of hallucinations associated with CBS.<sup>38</sup>

Risperidone, a drug used in the treatment of hallucinations associated with dementia, was successful in reducing hallucinations in one patient with CBS plus. Diazepam twice daily diminished visual hallucinations in one patient with true CBS, however, this patient also received individual and group psychotherapy and improved lighting.<sup>40</sup>

In contrast to these findings, it has been suggested that neuroleptic agents might lower seizure thresholds and increase the tendency to hallucinate.<sup>6</sup>

## Social management

Several groups have purported that the best form of rehabilitation for CBS is disclosure to a sympathetic professional, reassurance of the benign nature of the hallucinations, counselling and an explanation of the syndrome's causes, using the phantom limb analogy.<sup>14,16,18,19</sup> Patients are more likely to admit to experiencing visual hallucinations to a non-judgemental third party.<sup>16</sup> The possibility of hallucinations should also be discussed with those people who have not acknowledged their presence or who have not experienced them.<sup>14</sup>

Social isolation may be a contributing factor to visual hallucinations associated with CBS<sup>7,8</sup> but it may be the quality and not the quantity of social contact that is important. 'Loneliness, the subjective experience of missing significant (high-quality) interpersonal relationships', low extraversion and shyness could be contributing factors.<sup>7</sup>

To avoid misdiagnosis, Verstraten<sup>19</sup> advocated the referral of patients with visual hallucinations (apart from those with true CBS and those who are not distressed) to a psychologist for a structured interview and psychological testing. Patients in whom the diagnosis is likely to be something other than CBS can then be referred to other specialists.

Patients with true CBS could be given the opportunity to join a 'psycho-education group' (without having to undergo psychological testing), 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, concentrating on something else/looking for distraction, hitting the hallucination or shouting at the hallucination may reduce the length of the hallucinatory period.<sup>8,18</sup> The intervention strategy proposed by Verstraten may have merit but it has not been validated.

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## DISCUSSION

Although CBS was first described in 1769, there is still much to be learned about the condition. Accurate diagnosis is critical, as

an incorrect diagnosis could lead to sufferers being referred for inpatient psychiatric care, which may be very distressing and is unlikely to reduce or abolish the occurrence of hallucinations associated with CBS. Although awareness is growing, many medical professionals know little about the condition. In one study, it was found that proper diagnosis had been made in only one of 16 patients who had consulted a general medical practitioner about the hallucinations they were experiencing,<sup>18</sup> and it has been estimated that one to two per cent of individuals with CBS are referred for inappropriate inpatient psychiatric care.<sup>25</sup>

Many people do not report their hallucinations to medical practitioners or optometrists<sup>18</sup> because they believe that they may be thought of as mentally ill.<sup>25</sup> Thus, it is important for clinicians to inform those who are susceptible to CBS about the condition and to ask direct questions about the appearance of hallucinations, if CBS is suspected.

Many of the reports discussed in this review, particularly those on visual management, provide data from uncontrolled studies with small samples, therefore, it is difficult to reach any firm conclusion on effective management and further studies using larger samples and randomised controlled trials are required. Nevertheless, we believe that patients with suspected CBS should be referred initially to a low vision specialist, who may be able to reduce or alleviate hallucinatory activity by optimising visual function, recommending techniques, such as illumination modification and blinking and by providing information and reassurance. If there is a self-help group in the locality, the patient should be encouraged to attend. If appropriate, the patient should be considered for surgery, for example, cataract extraction. When optical or surgical intervention is either ineffective or inappropriate, referral to a specialist for pharmacological therapy can be instigated.

It is important for clinicians to learn more about CBS, so that they can recognise the symptoms and give sufferers appropriate information and advice. Discovering that true CBS is not related to mental

illness can often provide great relief for patients.<sup>18,25</sup>

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Author's address:

Dr F Eperjesi  
Neurosciences Research Institute  
School of Life and Health Sciences  
Aston University  
Birmingham B4 7ET  
UNITED KINGDOM