Charles Bonnet syndrome: a review

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Purpose of review

The aging of the population and the resultant increase in the number of patients with low vision due to age-related macular degeneration and other ocular diseases necessitate an increase in awareness of the Charles Bonnet syndrome among ophthalmic care providers.

Recent findings

The clinical features of Charles Bonnet syndrome have been described by several different authors as formed visual hallucinations due to disturbances of the visual system in patients who are otherwise mentally normal. Theories regarding the causes underlying the Charles Bonnet syndrome are multifaceted and offer insight into the function of the visual system. The incidence of the Charles Bonnet syndrome varies among different population groups, but is underdiagnosed in most settings. Recent case reports of treatment options involve varied pharmacologic interventions, but visual improvement and patient reassurance remain the mainstays of treatment.

Summary

As Charles Bonnet syndrome becomes more prevalent as the population ages, all physicians who care for low vision or elderly patients should be aware of its clinical characteristics and treatment options. Understanding of this syndrome by caregivers will lead to decreased anxiety among the patients who experience it. Further exploration of treatment options will be necessary in the future.

Keywords

Charles Bonnet syndrome, low vision, visual hallucinations

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Introduction

The Swiss philosopher and naturalist Charles Bonnet (1720–1792) first described the phenomenon of complex visual hallucinations in visually impaired, but otherwise psychologically normal patients in the 1760s. He first observed the phenomenon in his grandfather, who was visually impaired due to cataracts [1,2]. These hallucinations are usually of vivid, formed, and realistic objects or people and tend to recur. In the Charles Bonnet syndrome (CBS), patients are typically aware that these images are not real, although they may find them disturbing. However, the hallucinations may also fit logically into the visual scene and thus be indiscernible from real objects [3•].

Hallucinations are perceptions of unreal images, sounds, or smells in the absence of an external stimulus. In older patients, hallucinations can be associated with neurological diseases such as Alzheimer's dementia, Parkinson's disease, delirium, sensory and sleep deprivation, and psychosis [3•]. Release hallucinations also appear to originate from the external world, but are experienced when there is inadequate stimulus to the sensory system. The hallucinations of CBS are usually complex, not disturbing, and are typically only visual. Although CBS may be an early marker for dementia in the elderly, most patients are neurologically normal [4]. A lesion at any level of the visual system may lead to CBS. Patients typically have a loss of central visual acuity contributing to their symptoms, although this is not always the case. However, age-related macular degeneration (ARMD) with secondary decline in central vision is the most common of a variety of ocular diseases that may lead to CBS [3•].

Clinical characteristics

Vukicevic and Fitzmaurice [3•] described a typical patient with CBS: a woman in her late 70s with ARMD and hearing loss, who has a lower educational level and who has told a family member about the hallucinations. Although any patient with a lesion of the visual system is susceptible to experiencing hallucinations, risk factors for CBS include age in excess of 64 years, social isolation, lower cognitive function, a history of stroke, poor lighting, and worse bilateral visual acuity [5,6]. Careful screening with directed questioning of susceptible populations is essential in identifying patients with CBS.

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Although the character of the hallucinations can vary widely among patients, there are certain commonalities to the images. Among a population with CBS secondary to ARMD, Khan *et al.* [7•] found that the images tend to be straight ahead (85%), colored (72%), and to have moving parts (63%). People and geometric patterns are the most frequently perceived images, reported by 20 and 16% of patients, respectively. Although the hallucinations are classically purely visual, two patients with CBS and associated auditory hallucinations have been reported [8]. The hallucinations tend to occur in the area of visual field defect, even if the central visual acuity is relatively intact. An interesting case report [9] describes visual hallucinations occurring in the hemianopic hemifield of a patient who experienced an occipital lobe infarct with largely intact central visual acuity. Another report [10] of two patients who experienced CBS following sudden visual loss due to central retinal artery occlusion described the hallucinations as occurring within and at the borders of the recovered visual field. The fact that the hallucinations occurred when the patients were experiencing visual recovery suggests that fluctuation in vision may trigger the hallucinations of CBS. This is supported by reports of patients experiencing complex hallucinations within days following treatment of exudative ARMD with both intravitreal bevacizumab and photodynamic therapy [11,12].

In 85% of a population of patients with CBS secondary to ARMD, the onset of hallucinations correlated with acute loss of visual acuity [5]. However, the timing and frequency of the hallucinations can vary widely among patients with CBS. They may be episodic, periodic, or chronic. The hallucinations are usually daily or weekly and tend to occur upon awakening [3•]. Khan *et al.* [7•] report that the hallucinations occur daily in 34% of patients with CBS. Also, patients who have been experiencing their hallucinations chronically tend to hallucinate less frequently than those who have recently begun experiencing hallucinations. The duration of the hallucinations is usually several minutes (70%), but can be seconds (18%), or hours (12%) [3•]. Typically, there is a distinctive pattern to the timing and frequency of the hallucinations for any given patient.

The degree and complexity of the hallucinations also vary among patients. However, no association has been found between the complexity of the hallucinations and the severity of visual loss [7•]. Furthermore, progression of visual loss does not correlate with the complexity of the visual hallucinations [13]. Patients typically describe the nature of the hallucinations as benign. On questioning, 71% of patients with CBS secondary to ARMD described the experience as emotionally neutral [7•]. It is important to realize that although the hallucinations of CBS themselves are usually benign, patient's experience may not be. Among 30 patients with CBS, all claimed to have insight into the unreality of the hallucinations; however, 18 out of 30 (60%) first experienced confusion due to the images. One-third of the same patient population described a fear of impending insanity secondary to the hallucinations [14].

Although extensive testing of 14 patients with CBS did not reveal any relationship to psychiatric disease, coexisting neurologic disorders have been described in CBS patients [15]. Walker and Keys [16] describe a patient who presented with formed hallucinations secondary to macular disease who was subsequently found to have Lewy body disease and dementia. Although a vast majority of CBS patients are neurologically normal, older patients are prone to dementia-related visual hallucinations as well. Most commonly, the dementia is due to Lewy body disease or peduncular hallucinosis. It is essential to be aware of the possible dual diagnoses of CBS and early dementia, particularly in elderly patients who are susceptible to both disorders.

Prevalence

The prevalence of CBS varies widely between studies. Variable inclusion criteria, inconsistent depth of questioning, and reluctance of patients to admit having hallucinations all lent to these differences. However, studies of specific populations susceptible to CBS can guide the clinician as to the potential prevalence of the disorder within a clinical practice. In one study of patients with bilateral central scotomas due to ARMD and visual acuity in the better eye less than or equal to 20/80 [13], 40% (21/53) were diagnosed with CBS. The high prevalence in this study is likely due to the inclusion of patients experiencing simple visual hallucinations, which does not qualify as CBS according to most authors. Another study of patients with ARMD subdivided hallucinations into simple and complex and found a lower prevalence of CBS. Among 100 consecutive patients with ARMD treated with photodynamic therapy, 5% admitted to experiencing complex visual hallucinations consistent with CBS, whereas 15% experienced simple photopsias not consistent with CBS [12]. A case–control study of 100 consecutive patients with ARMD [5] found a 13% prevalence of CBS. A larger series of 360 cases of late ARMD identified 97 patients (27%) who had symptoms consistent with CBS. In this study, visual acuity worse than 6/36 (20/120) was associated with the presence of CBS [7•]. Another large study of patients more than 60 years old with multiple ophthalmic diagnoses resulting in visual acuity of 6/12 (20/40) or less found a prevalence of 17.5% (35 patients). When these 35 patients were compared with a randomly selected group of non-CBS controls over

the age of 60 years and with visual acuity of 6/12 or less, no difference between visual acuity, age, or sex was found [3•]. In another large study [6], the prevalence of CBS was found to be 11% in patients with acuities worse than 0.3 (20/67).

There is a high rate of patient nonreporting of visual hallucinations to care providers, largely due to a fear of being labeled with a psychiatric disorder. Vukicevic and Fitzmaurice [3•] found that whereas 21% of CBS patients had not reported their symptoms to anyone, 64% had mentioned them to a family member and 15% had told their ophthalmic care provider. Directed questioning by physicians to patients and their family members regarding hallucinations is essential for identifying patients with CBS.

Causes

The common characteristics among patients with CBS offer insight into the causes, and possible treatments, of the disorder. Two theories in particular are accepted as possible explanations of the pathogenesis of the hallucinations of CBS. The 'release theory' suggests that a lesion of the visual pathway results in abnormal signals being sent to the visual cortex. The anomalous signals, when combined with normal signals at the level of the visual association cortex, result in hallucinations. The 'deprivation theory' suggests that reduction in sensory input ('sub-threshold' visual input) leads to the production of spontaneous images from the visual association cortex, resulting in visual hallucinations [3•]. Menon *et al.* [17] describe the release phenomenon of the visual associative cortex as 'phantom images' akin to phantom limb pain. The sensory deprivation theory is supported by the increased likelihood of patients with CBS to have preexisting hearing loss and to be socially isolated. Further insight into the cause underlying CBS will hopefully lead to targeted biochemical treatments that decrease the frequency and duration of the symptoms.

Treatment

The mainstay of treatment for CBS is patient reassurance that the hallucinations are inherently benign and not the harbinger of psychiatric disease. The stress related to experiencing the hallucinations is usually due to concern over the underlying cause of the hallucinations and not necessarily the images themselves [3•]. Sympathy from a care provider and reassurance that the hallucinations are not a sign of underlying psychiatric disease led to increased patient comfort in 16 of 17 patients (94%) and is often all that is needed [14]. Additionally, increased social interaction and improved home lighting should be encouraged.

Along with patient reassurance and counseling, efforts to improve visual acuity should also be explored. A case report describes a 77-year-old patient who experienced formed visual hallucinations after his vision dropped to 20/400 due to a pigment epithelial detachment from ARMD. After treatment with pegaptanib and resolution of his pigment epithelial detachment, his vision improved to 20/50 with resolution of his hallucinatory symptoms [18]. Such cases suggest that increasing normal input to the visual association areas of the cerebral cortex can diminish the sub-threshold signals that cause visual hallucinations. Although low-vision therapy resulted in a decrease in the frequency of hallucinations in only three out of 11 patients with CBS, improvement in visual function may help patients cope with CBS [19].

Several case reports suggest that treatment with certain pharmacologic agents can be very effective in the alleviation of the hallucinations associated with CBS. The treatment of a man with CBS with the atypical antipsychotic olanzapine (5 mg/day) resulted in progressive resolution of the visual hallucinations over 7 days, at which point the medication was gradually withdrawn. The patient restarted therapy when the visual hallucinations recurred 3 months later and remained asymptomatic on continuous therapy of olanzapine 5 mg/day for 1 year [20]. Ukai *et al.* [21] describe a patient with CBS who remained symptom-free for 40 months on treatment with donepezil (a cholinesterase inhibitor). In a patient with CBS lasting 1 year due to visual loss to the hand motions level, treatment with the selective serotonin reuptake inhibitor venlafaxine resulted in the complete resolution of hallucinations in 4 days [22]. Additionally, one of our authors (J.B.S.) has found tricyclic antidepressants to be useful for treating CBS.

Conclusion

The first step in helping patients who are experiencing hallucinations due to vision loss is making the diagnosis of CBS. Directed questioning of susceptible patients is essential, as most patients are reluctant to admit hallucinations due to fear of being labeled with a psychiatric disease. In the elderly population, the clinician should be aware that CBS may be an early marker for dementia, which may necessitate a referral for specific evaluation and treatment.

A multidisciplinary approach is essential in managing and treating patients with CBS. The symptoms may resolve spontaneously, but this is often not the case. Improving visual function, addressing social isolation, and initiating pharmacological therapy are all important options to consider for CBS patients [17,23]. Physician awareness, empathy, and reassurance of the benign nature of CBS are key factors in helping these patients.

References and recommended reading

Papers of particular interest, published within the annual period of review, have been highlighted as:

- • of special interest
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Additional references related to this topic can also be found in the Current World Literature section in this issue (p. 226).

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