

Review Article

Navigating the Visual Distortions Associated with Charles Bonnet Syndrome

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A B S T R A C T

The term Charles Bonnet Syndrome (CBS) describes the visual hallucinations that arise from the brain adapting to a substantial loss of eyesight. Age-related macular degeneration and other eye diseases that impair vision are more common in the elderly than in any other age group.

Damage to the visual pathway causes visual hallucinations in people with CBS. These patients retain entire intellectual functioning along with partial or complete knowledge that the imaginations are not genuine, absence of psychosocial disorders, and absence of hallucinations impacting other sensory organs. The literature on neurology, geriatric medicine, and psychiatry has extensively recorded cases of CBS; however, optometry and ophthalmology have not provided as much material. Therefore, practising clinicians need to be more aware of the symptoms and signs linked with CBS. This review discusses the pathophysiology of CBS, alternative causes of visual hallucinations, and practical therapeutic techniques.

Keywords: Visual Hallucination, Charles Bonnet Syndrome, Impaired Vision, Visual Pathway, Neurobiology

Introduction

The disorder known as Charles Bonnet Syndrome (CBS), or "phantom vision," is characterised by visual hallucinations brought on by impairment to the visual pathway. Individuals with CBS do not have any other psychological disorders and are able to recognise that their visual hallucinations are not genuine.¹ In addition, the use of specific drugs, metabolic problems, psychological disorders, illicit drug usage, and neurological diseases can all result in visual hallucinations. Patients are reluctant to reveal that they experience visual hallucinations because they fear not receiving emergency care or being labelled as mentally unstable by friends, family, or medical professionals. The literature in the fields of neurology, geriatric medicine, and psychiatry has extensively reported this syndrome.

It has been suggested that sensory deprivation, social isolation, and cognitive dysfunction are the main causes of CBS. Due to a dread of confessing hallucinations and a fear of being classified as mentally handicapped, CBS is frequently overlooked until the geriatric age range.²

Patients who become aware of how fictitious their hallucinations are may experience depression due to their anxiety about becoming insane soon. It is necessary to have a thorough and comprehensive background in order to rule out hallucinations. The hallucinations may go away on their own, get better with vision, or the patients may get better at coping with social isolation.³ Figure 1. Reflects Comparison of a Normal Eye Lens with that of a CBS-Diagnosed Person.

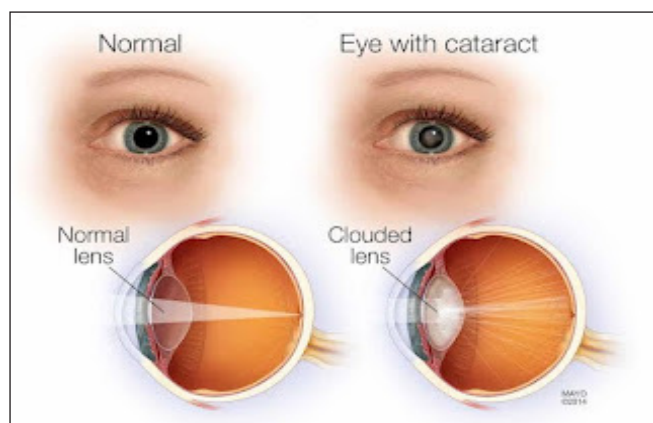


Figure 1. Comparison of a Normal Eye Lens with that of a CBS-Diagnosed Person

Symptoms

Hallucinations happen while the affected person is awake. They are often just visual in nature and unrelated to auditory (sound) hallucinations. Although such people may be first perplexed by visuals, they are usually aware that the hallucinations are not genuine.

Hallucinations related to CBS are usually not frightening, although they might be entertaining or bothersome. They occur without cognitive or psychiatric illness and come and go, frequently over months or years, with no apparent cause. Hallucinations are experienced by approximately one in five patients with retinal illnesses such as macular degeneration.⁴ These conditions can also affect patients with other macular diseases, ocular conditions such as glaucoma, and stroke.

Although it can happen at any age, CBS is more frequent among those who are 80 years of age or older. People who experience CBS may not necessarily have worsening eye conditions; in fact, visual hallucinations can occur in those with modest visual loss or small blind spots. Although the numbers may not accurately reflect the true prevalence, 10–30% of individuals with visual impairment involving both eyes are thought to be affected by CBS.⁵

Some people who experience hallucinations may believe they are “going crazy,” therefore, they may be afraid of what other people will think of them and decide not to tell anybody, even their doctors. Sometimes in the past, people were mistakenly admitted to acute mental health facilities before it was determined that their sight loss was the cause of their hallucinations. Over time, doctors have become more knowledgeable about CBS, and ophthalmologists should take into account the possibility that systemic disorders could be the cause of hallucinations. Making the right diagnosis of CBS can be greatly aided by the patient’s history.⁶

Epidemiology

Additional research is necessary to fully understand the incidence and prevalence of CBS. Studies and polls, however, suggest that the illness is not as rare as one may believe. Estimates of CBS prevalence vary greatly depending on sample size, ranging from approximately 1 to 10%.

A study that looked at CBS revealed that its total prevalence was 0.5%, with 0.8% of the sample having less visual acuity, 0.6% being elderly individuals, and 0.8% being in the category of both elderly people and those with poor visual acuity.⁷

A study that compared the prevalence of CBS in patients with macular disease and glaucoma who were undergoing visual rehabilitation revealed that the condition was more common in those with an upper degree of visual impairment or even more so in those who had lost all vision. The percentages were 39% and 20%, respectively.⁸

Age-wise, CBS can impact people of any age; however, it primarily affects the elderly. The usual age range for the problem, according to CBS data, is from old, which is the expected age range more than 70 years, in which pharmacological processes leading to visual conditions or loss of eyesight are expected to occur.

Nonetheless, research has indicated that a vast majority of CBS cases go unreported, primarily because people are afraid of receiving a mental health diagnosis; for example, a study in a retinal clinic revealed that just 12% of patients knew about CBS, indicating that a sizable portion of patients are ignorant of the condition. In contrast, a different study discovered that only 11–15% of elderly people with vision impairments reported experiencing visual hallucinations.

Additionally, children with a high rate of fast visual loss have been documented to have CBS.⁹ The elderly are also more likely to experience profound sight loss, according to reports. De Mossier saw a preponderance of men,¹⁰ other research revealed a preponderance of women,¹¹ and many other investigations revealed no sex bias.^{12,13}

Pathophysiology

Any factor that results in visual loss is the initial step in the pathophysiology of CBS. Frequently seen conditions, which can impact any aspect of the visual pathway, are age-related macular degeneration, diabetic retinopathy, cataract, glaucoma, and cerebral infarctions that impact the visual cortex.

The most widely recognised theory now in use to explain the visual hallucinations associated with CBS is the disinhibition of cortical regions that impact vision as a result of visual sensory deafferentation.¹¹

In the end, this disinhibition causes these vision-related

regions to fire spontaneously, which causes hallucinations. In the end, this disinhibition causes these vision-related regions to fire spontaneously, which results in hallucinations. Research bolstering this notion has demonstrated via neuroimaging that, in individuals who matched diagnostic criteria for visual hallucinations, there is increased spontaneous activity and functional connectivity within the visual cortex and associated perceptual networks even in the absence of external stimuli.

Additionally, MRI demonstrated a correlation between the hallucinations and the activities of the firing cortical region involved. In the end, this disinhibition causes these vision-related regions to fire spontaneously, which results in hallucinations. Research supporting this notion has demonstrated through neuroimaging that individuals who meet the diagnostic criteria for visual hallucinations exhibit increased spontaneous activity and functional connectivity within the visual cortex and associated perceptual networks, even in the absence of external stimuli. As a result of the mechanism by which hallucinations are generated and interpreted, individuals with congenital blindness do not exhibit Charles Bonnet Syndrome (CBS).¹² This is because CBS arises from visual cortical disinhibition following sensory deprivation, a process that requires prior visual experience — something absent in those who are congenitally blind.

Aetiology

- Diabetic retinopathy
- Macular degeneration associated with ageing
- Cerebrovascular accidents
- Glaucoma
- Degeneration of the macular pigment
- Cataract
- High degree of nearsightedness (high myopia)
- Retinitis pigmentosa
- Optic nerve injury
- Occlusion of the retinal vein
- Occlusion of the central retinal arteries
- Stroke in the occipital region
- Temporal arteritis

A phantom limb pain explanation is the one put forth to explain CBS. The hypothesis of phantom limb pain explains why a person may experience pain even after losing a limb. Despite having the limb amputated, the patient still experiences discomfort. Similarly, even if they are blind, people with CBS may experience visual encounters.¹³

It is imperative to identify the aetiology of the visual hallucination to ensure proper management. Figure 2 demonstrates visual Delusions and RAY Vision as a Tool.

The prevalence of CBS was assessed by Crane et al. Out of the 284 participants in their study, 57.7% had diabetes,

19.7% had retinal disorders, 15.4% had neuropathic illness, 12.32% and had additional eye conditions (cataract, amblyopia, cerebrovascular accident, corneal scar, nystagmus, pseudophakia bullous keratopathy, and uveitis). Subedi et al. reported that CBS was present in patients with advanced retinitis pigmentosa.¹⁴

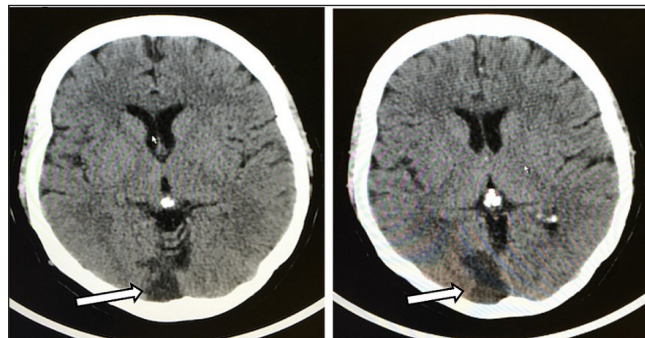


Figure 2. Visual Delusions and RAY Vision as a Tool Neurologic Disorder

Parkinson's Disease: The pathophysiology of Parkinson's disease extends beyond the substantia nigra and includes significant changes to the brainstem and many neurotransmitter systems. Patients suffering from this disease have a marked neuronal loss in the brain stem. These patients have abnormalities in the visual pathway linked to central and retinal dopamine deficiencies. Dopamine levels are raised by levodopa, which should enhance the performance of the visual system. Visual hallucinations in these patients are related to reduced eyesight, cognitive decline, dopaminergic drugs, or anticholinergic drugs.¹⁵

Manford and Andermann state that the first reports of visual hallucinations occurred after anticholinergics, levodopa, and dopaminergic agonists were started as a treatment for Parkinson's disease. More precisely, after approximately ten years of levodopa therapy, about ninety percent of individuals began experiencing hallucinations. It was formerly believed that the dosage of levodopa was directly associated with hallucinations. Levodopa's direct toxic effect on vision, however, does not appear to be the primary source of hallucinations. ninety Visual hallucinations, which are linked to vivid dreams, sleep difficulties and shifts in arousal, typically happen towards the end of the day. Patients with Parkinson's disease continue to realise that their hallucinations are not genuine.¹⁶

Pathological Hallucinoses: There are many clinical parallels between CBS and Peduncular hallucinosis. The underlying cause of Peduncular hallucinosis is veins. Damage to the thalamus or midbrain might result in complex visual hallucinations. The midbrain and its environs are included in the term peduncular, which does not simply refer to brain peduncles. Other pathologies of the central nervous system that have been linked to Pendulum hallucinosis include

vascular and infectious lesions of the midbrain, the pontine, and thalamus; local subarachnoid haemorrhage; basilar vascular hypoplasia; basilar migraine; and compression brought on by local and distal malignancies. It is also seen in patients following angiography and surgery.¹⁷ Hallucinations of people, animals, landscapes, twisted or unsightly faces, repeating patterns, and Lilliputian creatures are common in these patients. "Lilliputian hallucinations" are visual phenomena that happen when the hallucinations are quite small.¹⁸

Peduncular hallucinosis usually manifests several days following the infarct. To distinguish it from CBS, each hallucination episode might be tactile or auditory and last anywhere from a few minutes to several hours. People with peduncular hallucinosis may have hallucinations for several weeks to years. These people are still aware that the hallucinations are imaginary.¹⁹ The hallucinations stop when the underlying central nervous system diseases are treated. Therefore, radiographic screening for central nervous system illnesses is essential to rule out peduncular hallucinosis.¹⁹

In addition to the cerebral and visual symptoms of multiple sclerosis, there have been reports of the development of visual hallucinations in multiple sclerosis (CBS). These patients usually experience some cognitive impairment as a result of the disease.

Causes of CBS

Visual impairment is strongly associated with the development of Charles Bonnet Syndrome (CBS). When vision deteriorates or blindness occurs, the brain receives reduced sensory input from the eyes. In response, it may compensate by generating spontaneous visual activity, which manifests as hallucinations.²⁰ However, not all individuals who are blind develop CBS. The likelihood of its occurrence increases under certain conditions, such as:

- Sudden or rapid loss of vision
- Bilateral (both-eye) vision loss
- Presence of underlying ocular disorders such as cataract or age-related macular degeneration

Importantly, CBS is not associated with dementia or psychiatric disorders.²¹

Individuals affected by CBS typically report a wide range of visual hallucinations, including:

- Recurring geometric patterns, dots, or lines
- Complex scenes featuring landscapes such as mountains or waterfalls
- People, animals, or insects
- Figures dressed in historical or elaborate costumes
- Imaginary or fantastical creatures, such as dragons

Diagnosis of CBS

Diagnosis of Charles Bonnet Syndrome (CBS): There is currently no specific diagnostic test to confirm Charles Bonnet Syndrome (CBS). Diagnosis is primarily clinical and relies on a detailed medical history and evaluation by a healthcare professional. It is essential to rule out other possible causes of visual hallucinations before confirming CBS. These include:

- Adverse effects of certain medications
- Underlying mental health disorders
- Neurological or structural brain abnormalities

A diagnosis of CBS can be established when an individual experiences visual hallucinations in the presence of visual impairment, without evidence of the following conditions:¹⁵

- Peduncular hallucinosis
- Levodopa-induced hallucinations
- Hypnagogic or hypnopompic hallucinations
- Major depressive disorder
- Epileptic seizures
- Alzheimer's disease or other dementias
- Migraine aura or migraine coma
- Neurodegenerative disorders such as Parkinson's disease or Lewy body dementia
- Delirium, substance (alcohol or drug) withdrawal, or metabolic encephalopathy

Treatment

CBS has no known cure or effective medical treatment, yet there are coping mechanisms and management strategies that can be employed.

Coping Mechanism

Patients experiencing visual hallucinations associated with Charles Bonnet Syndrome (CBS) have reported that several behavioral techniques can help reduce or temporarily eliminate hallucinations.²² These strategies aim to stimulate visual and cognitive pathways, thereby interrupting the abnormal visual activity. Effective methods include:

- When a hallucination begins, look from side to side every fifteen seconds without moving the head. This maneuver may need to be repeated four or five times, but if it does not help, it should not be continued beyond five attempts.
- Blink several times to reset the visual field.
- Focus directly on the hallucination to help it fade.
- Attempt to interact mentally or verbally with the hallucinated image.
- Rotate the head gently from one side to the other, or towards each shoulder in turn.
- Change your position by moving around the room or shifting to a different location.

- Illuminate the surroundings—for example, by shining a flashlight upward from beneath the chin (avoiding direct exposure to the eyes).
- Adjust ambient lighting to suit the activity or environment, as both excessive brightness and dimness can exacerbate hallucinations.

Learning

Patients with CBS frequently suffer from worry and anxiety, which is related to their confusion about the cause and significance of the hallucinations. This is particularly true if they have not previously been made aware of this possible side effect of their sight loss.

As a result, it is crucial to educate patients, family members, and medical professionals about CBS. Enhancing patients' quality of life can be achieved by educating them and then providing them with coping mechanisms.²³

Pharmacotherapy

Medications are usually prescribed for serious illnesses only, such as those accompanied by unsettling or persistent hallucinations. Although the effectiveness of antipsychotics has been found to be inconsistent, because of their less dangerous side effects, atypical antipsychotics, such as low doses of quetiapine or olanzapine, are advised, especially for the older demographic that CBS tends to affect.

Among the other drugs with good effectiveness and a few negative effects are cholinesterase inhibitors, such as donepezil. Antiepileptic drugs such as valproate, carbamazepine, gabapentin, and clonazepam have been shown to be beneficial anecdotally. Last but not least, prokinetic drugs like tiapride and antidepressants like venlafaxine and escitalopram have shown promise, as shown in a limited case series.²⁴

Psychological Therapy

The promotion of various tactics, such as hypnosis, relaxation training, diversion, cognitive remodelling, and psychological therapy for phantom experiences, has helped lessen the unpleasant and troublesome impacts of visual hallucination.²⁵

Conclusion

There is a greater chance that more patients will present with CBS in the future as the population ages and more people have impaired vision from a variety of ocular disorders. Therefore, it is vital to familiarise the medical fraternity with the syndrome so that primary care optometrists can correctly diagnose and treat CBS in their patients. Treatment for the underlying eye condition, as well as fitting the patient with the proper vision-correcting devices, is a part of managing these hallucinations.

Declaration of Generative AI and AI-Assisted Technologies in the Writing Process:

During the preparation of this thesis, generative AI and AI-assisted technologies (OpenAI's ChatGPT) were utilized to support tasks such as language refinement, grammar correction, reference formatting, paraphrasing, and improvement of academic readability. All conceptualization, data interpretation, experimental design, and conclusions were entirely developed by the author. The AI tool was used solely to enhance clarity and presentation without influencing the scientific content or integrity of the work.

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