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# Hallucinations Experienced by visually impaired: Charles Bonnet Syndrome

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***Abstract***
*Charles Bonnet Syndrome is a condition where visual hallucinations occur as a result of damage along the visual pathway. Patients with Charles Bonnet Syndrome maintain partial or full insight that the hallucinations are not real, absence of psychological conditions, and absence of hallucinations affecting other sensory modalities, while maintaining intact intellectual functioning. Charles Bonnet Syndrome has been well documented in neurologic, geriatric medicine, and psychiatric literature, but there is lack of information in optometric and ophthalmologic literature. Therefore, increased awareness of signs and symptoms associated with Charles Bonnet Syndrome is required among practicing clinicians. This review of the literature will also identify other etiologies of visual hallucinations, pathophysiology of Charles Bonnet Syndrome, and effective management strategies.*
*Charles Bonnet Syndrome, also known as "phantom vision," is a condition where visual hallucinations occur as a result of damage along the visual pathway. People experiencing Charles Bonnet Syndrome have intact cognition, insight that the visual hallucinations are not real, and absence of other psychological conditions.[1] Visual hallucinations may also occur in neurological diseases, metabolic disorders, psychiatric conditions, illicit drug use, and use of certain medications. Patients do not willingly disclose that they experience visual hallucinations for fear of not being taken seriously or fear of being labeled as mentally unstable by their loved ones and/or health care providers. Charles Bonnet Syndrome has been well documented in the neurologic, geriatric medicine, and psychiatric literature. There is lack of information in the optometric and ophthalmologic literature; therefore, it is still poorly understood by eye care professionals. This review of the literature aims to increase awareness about Charles Bonnet Syndrome, improve recognition of the symptoms, identify the condition, and offer management strategies among practicing clinicians.*

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**History**

In 1760, a Swiss scientist named Charles Bonnet, who was well known for his contributions to botany and philosophy, first described visual hallucinations associated with Charles Bonnet Syndrome. Bonnet documented the complex visual hallucinations experienced by his 90-year-old grandfather, Charles Lullin, and published his findings in an essay.1 His grandfather underwent cataract surgery in both eyes. The bilateral cataract surgery initially improved his vision, but his vision deteriorated over time. The visual hallucinations occurred shortly after his vision loss. Charles Lullin's hallucinations included detailed figures of men, women, birds, animals, buildings, tapestries, and carriages with variations in size, shape, and place.1,2 He knew that these visual hallucinations were not real, but he was uncertain why he was experiencing them. He was in good overall health without any psychiatric disorders.

Ironically, Charles Bonnet suffered from an unknown cause of painful vision loss at 20 years of age. The vision loss progressed to severe visual impairment by the age of 40. He also experienced visual hallucinations. However, it was not until 1967 that another Swiss scientist, George De Morsier, coined the term, "Charles Bonnet Syndrome." De Morsier was known well for his contributions in neurodevelopmental disorders.

**PREVALENCE**

The prevalence of Charles Bonnet Syndrome varies and ranges from 0.4% up to 30%, depending on the study.[1,3–6] Khan et al.[4] noted that the incidence of Charles Bonnet Syndrome was statistically higher in subjects with worse visual acuity. This finding demonstrated worse vision is associated to some degree with increased risk of experiencing visual hallucinations.[4]

There are several reasons why variability in the prevalence rates of Charles Bonnet Syndrome exist. First, there is no universal definition of Charles Bonnet Syndrome agreed upon by all healthcare professionals. Second, the diagnosis of Charles Bonnet Syndrome is made across different disciplines including optometry,

ophthalmology, geriatric medicine, psychiatry, and neurology. Third, many people are hesitant to report that they experience visual hallucinations for fear of being labeled as mentally unstable.[1,7–9] As the geriatric population increases, the amount of acquired vision loss will also increase. As a result, there will be an increase in those affected by Charles Bonnet Syndrome. As health care providers, we need to understand and recognize this condition so these individuals can be properly managed. In a study by Gilmour et al., only 9% of people experiencing Charles Bonnet Syndrome sought medical advice. Disappointingly, only half of them received an explanation about Charles Bonnet Syndrome.

In another study, 15.2% of the participants informed their eye health professional about the visual hallucinations. Cox and Ffytche investigated 492 people with Charles Bonnet Syndrome. There was a trend association between the quality of information provided by their medical professional and whether a negative outcome was experienced. Forty-seven percent of the people with Charles Bonnet Syndrome were not given a clear explanation and experienced negative outcomes whereas 36% of people with Charles Bonnet Syndrome were given a clear explanation. They also found one third of the medical professionals were either uncertain or unaware about Charles Bonnet Syndrome.

**DEFINITIONS**

There are many definitions that have been used to classify Charles Bonnet Syndrome, but there has been no consensus on which definition is most accurate. In 1935, According to De Morsier's definition,10 Charles Bonnet Syndrome implied a localized neurodegeneration, such as is evident in Parkinson's disease, Alzheimer's disease, and peduncular hallucinosis. He strongly believed Charles Bonnet Syndrome occurred in the elderly with normal cognition and without correlation to ocular pathology. The fact that Charles Bonnet's grandfather had impaired vision was merely a coincidence that ocular pathology is common in the elderly.10 De Morsier was not able to provide evidence to confirm his hypothesis.

In 1936, L'Hermitte and de Ajuriaguerra's definition implied visual hallucinations involve thalamic lesions in addition to ocular pathology.10 They did not agree with De Morsier's definition that Charles Bonnet Syndrome occurred in the absence of ocular pathology. L'Hermitte and de Ajuriaguerra's definition coincided with Bonnet's original description of visual hallucinations experienced by his grandfather.

Gold and Rabins' definition[10] (1989) is most commonly used in the psychiatric literature. They describe the visual hallucinations in Charles Bonnet Syndrome to be stereotyped, formed, complex, persistent, or repetitive in nature.[10] Further, they suggest that those with Charles Bonnet Syndrome are aware that the hallucinations are not real. In their definition, there are no delusions or hallucinations affecting other sensory modalities. Ocular pathology is not part of their diagnostic criteria.

Ophthalmologists and neurologists use Charles Bonnet Syndrome to describe visual hallucinations that occur as a result of ocular pathology or visual pathway disease. The type of hallucinations and age of the patient are of secondary importance.[12] The definition used by ophthalmologists and neurologists supports Bonnet's original description of the condition. An important consideration about the ophthalmologically defined Charles Bonnet Syndrome is that it carries an inherent ambiguity: Was ocular pathology or loss of acuity the contributing factor for the visual hallucinations? Diagnostic criteria for Charles Bonnet Syndrome still remains controversial.

**CHARACTERISTICS OF CHARLES BONNET SYNDROME**

Visual hallucinations experienced by Charles Bonnet Syndrome patients are typically simple or complex in nature, although a full spectrum of hallucinations can occur. Simple visual hallucinations, which are sometimes described as elementary or formed, are composed of photopsias, simple shapes, grid-like patterns, and branching patterns. Complex visual hallucinations are made up of vivid and complicated images of people, faces, vehicles, animals, flowers, trees, plants, and miniature images of people and objects.

The visual hallucination episodes last seconds to a few hours, with recurrent episodes of visual hallucinations occurring for days to years. A majority of the visual hallucinations are strange or bizarre to the patients, but they are seldom disturbing. Generally, people experiencing visual hallucinations remain neutral about the hallucinations.

In a study done in the United Kingdom, 38% of the 492 subjects described the visual hallucinations as frightening, terrifying, and startling during the initial onset of them. Over time, their emotions towards the hallucinations decreased to 8%.56 They also found 60% of subjects reporting the visual hallucinations did not affect their lives, 33% of subjects felt it had a negative effect on their lives, and only 7% felt it had a pleasant effect on their lives.

A Canadian study mirrors Cox et al.'s results citing 63% individuals experienced a neutral reaction to the hallucinations, 34% had a negative reaction, and 6% had a positive response to them.48 These recent findings demonstrate the prevalence of negative reactions to visual hallucinations are higher than previously reported by Holroyd and Rabins.

In 2008, Vukicevic and Fitzmaurice found 51.5% of their participants report mild stress compared to 30% of participants who report severe stress as a result of visual hallucinations. The stress was not a result of the images seen, rather it was primarily due to concern about the underlying cause of the hallucinations.

In case report, Vukicevic demonstrated how a stressful life event can change the nature of hallucinations experienced in Charles Bonnet Syndrome from non-distressing to distressing. An 80-year-old woman with diagnosed age-related macular degeneration and closed-angle glaucoma had been experiencing Charles Bonnet Syndrome for 4 years. Her hallucinations became atypical on February 7, 2009. This date was marked by intense heat and bushfires in southern Australia known as "Black Saturday." She lived in an area affected by the fires and evacuated from her house on that day. The patient likely suffered from acute stress disorder because she had a history of fire-related trauma. The change in nature of her hallucinations may represent a relationship between Charles Bonnet Syndrome and acute or posttraumatic stress disorder.

**Charles Bonnet Syndrome and Visual Acuity**

Madill and colleagues studied the occurrence of Charles Bonnet Syndrome in four patients with advanced glaucoma.[12] They challenged previous authors who stated that acuity loss is required for ophthalmologically defined Charles Bonnet Syndrome. In these advanced glaucoma patients, visual acuity ranged from 20/20 to 20/40 monocularly with extensive visual field defects. No hallucinations occurred in other sensory modalities, and all maintained insight that the hallucinations were not real. The results of Madill et al.'s case study coincide with the ophthalmological definition of Charles Bonnet Syndrome emphasizing ocular pathology or visual pathway disease as a cause of the visual hallucinations. It is important to understand this case series emphasizes visual acuity loss is not a requirement for Charles Bonnet Syndrome visual hallucinations to occur.

With that said, an association between age-related macular degeneration (AMD) and colored visual hallucinations has been proposed. Signals related to color vision pass through the parvocellular layers of the lateral genicular nucleus (LGN) and are transmitted along the ventral color pathway. In patients with macular degeneration, there is degeneration throughout the ventral color pathway.15 Damage to cone photoreceptors in the macula creates a lack of visual input to the color areas of the visual association cortex. Because of reduced visual input to the visual association cortex, endogenous activation within the color area results in colored hallucinations. Functional magnetic resonance imaging (fMRI) provides support for this theory, as the color area of the visual association cortex remained hyperactive in Charles Bonnet Syndrome patients with macular degeneration. Because of the physiological deafferentation with ganglion cell loss in glaucoma, they can experience Charles Bonnet Syndrome without an acuity loss, whereas in AMD, there is loss of central retinal ganglion cells, which causes Charles Bonnet Syndrome to occur with an acuity loss.12 Therefore, patients with significant ocular disease and preserved visual acuity may still be at risk for developing Charles Bonnet Syndrome. In a study evaluating patients only with AMD, their extent of vision loss did not predict which patients would experience visual hallucinations. The investigators also found that the extent of vision loss did not determine the complexity of visual hallucinations experienced.57 Gilmour and colleagues found no statistical significance in patients with dry or wet AMD and likelihood of having visual hallucinations. Patients with both types of AMD experienced Charles Bonnet Syndrome.

**No Age Limit for Charles Bonnet Syndrome**

According to Khan et al.4 and Menon's studies,16 there were no increased risks of Charles Bonnet Syndrome with increased age. Although Charles Bonnet Syndrome affects geriatric populations because of increased incidence of eye disease in the elderly, it can affect all ages after vision loss.7 In the original definitions of Charles Bonnet Syndrome, there have been no references to age being required as part of the inclusion criteria. Although very few studies of Charles Bonnet Syndrome in children are found in the literature, Schwartz and Vahgei reported two cases of Charles Bonnet Syndrome that occurred in children after profound vision loss.17 Both children had a diagnosis of cone-rod dystrophy accompanied by vivid and complicated visual hallucinations.

The children's visual hallucinations were very similar to hallucinations described by adults with the condition.17 The complicated visual hallucinations included flashing lights, people, faces, wolves, snowballs, ballerina, houses, and colored balls. No other sensory modalities were affected. This study highlighted the important concept that age is not a criterion for the diagnosis of Charles Bonnet Syndrome.17 Visual hallucinations have not been reported by those with congenital blindness. In fact, Charles Bonnet Syndrome has only been reported as being partially related to visual impairment.

**ETIOLOGIES OF VISUAL HALLUCINATIONS**

Determining the etiology responsible for the visual hallucinations is essential to implement the appropriate management. Table 1 outlines a sampling of the common etiologies of visual hallucinations.

| Neurologic disorders | Psychiatric disorders | Illicit drugs | Metabolic disorders | Miscellaneous conditions | Charles Bonnet Syndrome | Prescription medicines |
|---|---|---|---|---|---|---|
| Intracranial tumors | Schizophrenia | LSD | Drug withdrawal | Hypnagogic hallucinations (transitional state between waking and sleeping) | Macular diseases | See Appendix A |
| Peduncular hallucinosis | Delirium | Psilocin | Alcohol withdrawal | Hypnopompic hallucinations (transitional state between sleeping and waking) | Retinal diseases | Medication side effects |
| Alzheimer disease | Acute psychoses | Psilocybin | Cardiopulmonary insufficiency | Narcolepsy-cataplexy syndrome | Optic nerve disease | |
| Aneurysms | Bipolar disorder | PCP | Uremia | Sleep deprivation | Central retinal artery occlusion | |
| Seizure disorders | Depression | Angel dust | Hepatic disease | Food deprivation | Central retinal vein occlusion | |
| Lewy body dementia | | Mescaline | Endocrine disturbances | Water deprivation | Field deficits without visual acuity loss | |
| Encephalitis | | Cannabis | Vitamin deficiencies | Prolonged social isolation | | |
| Meningitis | | Amphetamines | Inflammatory disease | Excessive fatigue | | |
| Head trauma | | Methylphenidate | Infectious disease | Excessive stress | | |
| Infarcts | | Cocaine | | | | |
| Migraines | | | | | | |
| Epilepsy | | | | | | |

**Table1**: Ethiologies of visual hallucinations

**Ocular Pathologies**

Crane et al. evaluated the prevalence of Charles Bonnet Syndrome. In their study of 284 individuals, 57.7% had macular disease, 19.7% had retinal disease, 15.4% had neuropathic disease, and 7% had ocular disease of other etiologies (amblyopia, cataract, central retinal artery occlusion, central retinal vein occlusion, cerebrovascular accident, corneal scar, nystagmus, pseudophakic bullous keratopathy, and uveitis).52 O'Hare et al. found 37% individuals with advanced retinitis pigmentosa experienced Charles Bonnet Syndrome.

**Neurologic Disorders**

**Parkinson's Disease**

The pathology of Parkinson's disease extends beyond the substantia nigra to include widespread brainstem changes and various neurotransmitter systems. There is significant loss of neurons in the brainstem of Parkinson's disease patients. In these patients, there are defects in the visual pathway associated with dopamine deficiency in the retina and central pathways. Levodopa increases the dopamine levels, which should also improve the visual system's function.19 In those with Parkinson's disease, visual hallucinations are attributed to decreased vision, cognitive impairment, dopaminergic medications, or anticholinergic medications.

According to Manford and Andermann in 1998, visual hallucinations were not reported until after Parkinson's disease treatment was initiated with anticholinergics, Levodopa, and dopaminergic agonists. More specifically, in patients who took Levodopa, their hallucinations began after 10 years of using the medication. It was thought that hallucinations were directly related to the dosage of Levodopa. However, a direct toxic effect of Levodopa on vision seems unlikely to cause hallucinations.19 Visual hallucinations usually occur towards the end of day, associated with vivid dreams, disturbances in sleep, and changes in arousal. Parkinson's disease patients maintain insight that the hallucinations are not real.

According to Biousse and colleagues (2004), one fourth of 30 Parkinson's disease patients experienced complex visual hallucinations.[20] Some authors believe visual hallucinations in Parkinson's disease indicate the development of dementia. However, the Biousse study found no dementia after 2 years of follow-up.

**Peduncular Hallucinosis**

Peduncular hallucinosis shows the closest clinical overlap with Charles Bonnet Syndrome.22 The underlying etiology of peduncular hallucinosis is vascular in nature. Complex visual hallucinations occur as a result of lesions in the midbrain and/or thalamus. The term "peduncular" does not only imply cerebral peduncles but includes the midbrain and surrounding area. Peduncular hallucinosis has been associated with other central nervous system pathologies, including vascular and infectious pontine, midbrain, and thalamic lesions, local subarachnoid hemorrhage, compression by local and distal tumors, basilar vascular hypoplasia, basilar migraine, and after surgical and angiographic interventions.22 Common hallucinations experienced by these individuals include people, animals, landscapes, grotesque and deformed faces, repeated patterns, and Lilliputian hallucinations. Lilliputian hallucinations are visual phenomena in which the hallucinations are miniature in size.

Peduncular hallucinosis typically occurs a few days after the infarct. Each hallucination episode will last for a few minutes to several hours and can be accompanied by tactile or auditory hallucinations differentiating it from Charles Bonnet Syndrome. Patients with peduncular hallucinosis may experience hallucinations for several weeks to years. These patients do maintain insight that the hallucinations are not real.19 Treating the underlying central nervous system pathologies will resolve the hallucinations. Therefore, it is important to screen radiologically for central nervous system disease to rule out peduncular hallucinosis.

**Multiple Sclerosis**

With the ocular and cerebral manifestations of multiple sclerosis, there are reports of Charles Bonnet Syndrome visual hallucinations that can occur. These patients will usually have some cognitive impairment due to the disease. According to Chen and colleagues, a woman with multiple sclerosis lost vision from optic neuritis and began experiencing complex and vivid visual hallucinations.9 In another case, a woman with multiple sclerosis noticed that her visual hallucinations disappeared once she regained vision.23 These case studies highlight that dynamic changes in visual acuity may play a role in eliciting Charles Bonnet Syndrome visual hallucinations, although it has not been proven.

**Epilepsy**

Visual hallucinations associated with epilepsy are different from other hallucinatory events. These hallucinations are brief, simple, and tend to be associated with seizure manifestations. Intracranial electroencephalograph and direct cortical stimulation experiments demonstrate excitation of visual cortical areas. A hallmark sign of epileptic hallucinations is excitation in the visual association cortex.19 Complicated visual hallucinations that occur in epilepsy involve the posterior parietal or temporal association cortex.

**Conditions Affecting Blood Supply**

**Migraines**

Auras before migraine headaches have been well recognized as a cause of simple visual hallucinations. It is rare to experience complex visual hallucinations with migraines. Investigators suggest that depression of cortical activity, in conjunction with changes in blood flow, are responsible for generation of migraine auras. The most common symptoms reported by patients with visual auras preceding migraines include photopsias and simple shapes and patterns, along with very bright and colorful images.7 Ocular migraines without headaches are encountered frequently in eye care practices. Therefore, practitioners must distinguish between visual hallucinations related to ocular migraines, typical migraines, and Charles Bonnet Syndrome to appropriately manage the visual hallucinations.

**Infarcts**

In cases of infarcts causing visual hallucinations, ischemia is the underlying etiology of cortical lesions. The onset of visual hallucinations occurs days to weeks after the ischemic event. These hallucinations have a typical duration of days to weeks, but the frequency may persist over time. The majority of complicated visual hallucinations, which are reported after stroke, are due to occipital infarcts, rather than middle cerebral artery infarcts.19 This is consistent with the hypothesis that epilepsy activates the visual association cortex directly. However, strokes stimulate the visual association cortex from a distance. Possible explanations for how strokes affect the visual association cortex include (1) loss of direct cortico-cortical inputs and (2) loss of striate cortex control of thalamic inputs to visual association cortices.

**Regional Cerebral Blood Flow**

A study evaluated regional cerebral blood flow using brain single-photon emission computed tomography (SPECT) in patients experiencing Charles Bonnet Syndrome visual hallucinations. There was an increased perfusion in regional cerebral blood flow in the regions of the lateral temporal cortex, striatum, and thalamus in patients who experienced visual hallucinations (Fig. 1–3). It is suggested that people with vision loss have increased cortical activation in the areas of the lateral temporal cortex, striatum, and thalamus contributing to development of visual hallucinations.

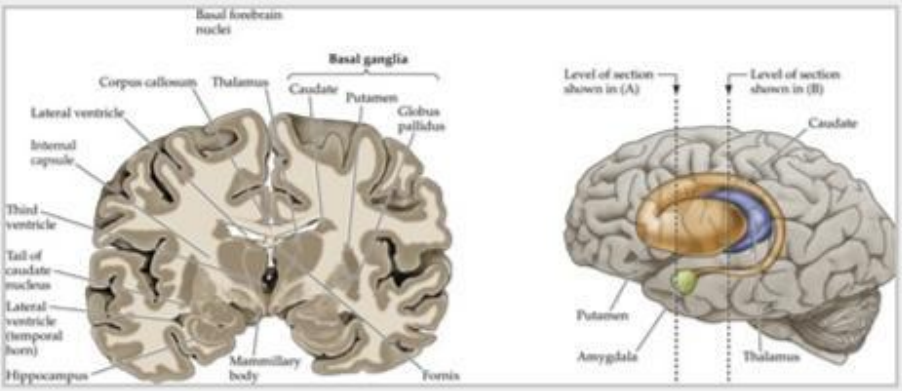


**FIGURE 1:** Diagram identifying the thalamus and striatum. Reproduced with permission from Purves DL, et al., eds. Neuroscience, 4th ed. Sunderland, MA: Sinauer; 2008.

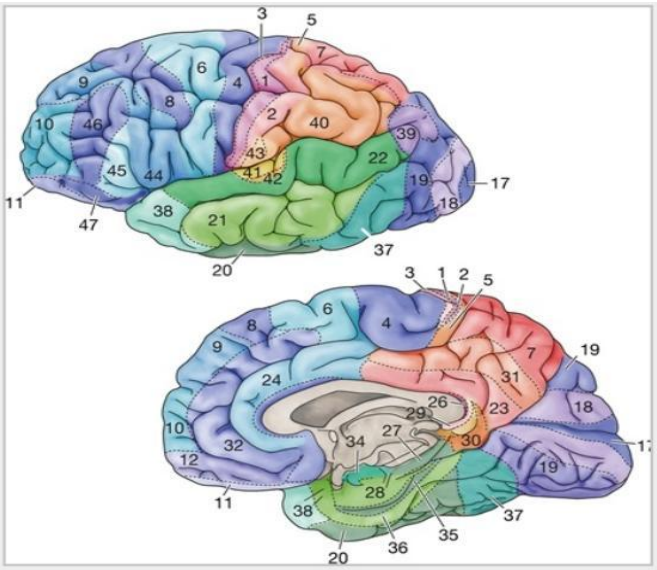


**FIGURE 2:** Visual cortical areas, including Brodmann Area 17 (primary visual cortex), Brodmann Area 18 (secondary visual cortex), and Brodmann Areas 19 and 37 (visual association cortices). Reproduced with permission from Purves DL, et al., eds. Neuroscience, 4th ed. Sunderland, MA: Sinauer; 2008.

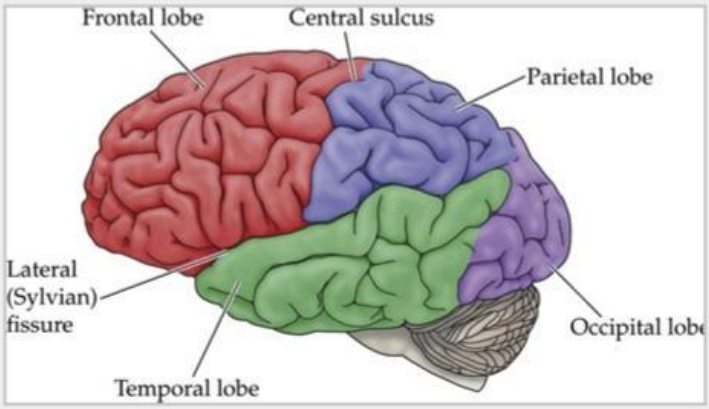


**FIGURE 3:** Diagram outlining the ventral occipitotemporal cortex (also known as the ventral occipital lobe). Reproduced with permission from Purves DL, et al., eds. Neuroscience, 4th ed. Sunderland, MA: Sinauer; 2008.

**PRESCRIPTION MEDICATIONS**

Prescription medications can have a side effect of visual hallucinations by mechanisms that are still unknown.19,26,27 Appendix A includes a comprehensive list of these medications (available at https://links.lww.com/OPX/A252).26,27 Alpha-2 agonists such as brimonidine tartrate have been shown to cause systemic and neuropsychiatric phenomena, which may induce Charles Bonnet Syndrome .28 Because it is a well-known lipophilic agent, it has the ability to penetrate the blood–brain barrier and cause systemic side effects such as visual and auditory hallucinations, depression, confusion, and anxiety in adults.29,44 In children, it can cause adverse effects such as coma, hypotension, bradycardia, hypotony, and hypothermia.44 The study by Tomsak et al.28 reported four primary open-angle glaucoma subjects experienced visual hallucinations within 5 days to 2.5 months after starting brimonidine tartrate therapy.28 The authors found the visual hallucinations resolved after discontinuation of brimonidine tartrate.28 A case report by Garcia-Catalan et al. describes an 81-year-old woman with pseudoexfoliation glaucoma and age-related macular degeneration developed visual hallucinations after 1 month of using brimonidine with complete resolution of the hallucinations after suspending its use.44 However, Rahman et al. concluded visual hallucinations are a rare side effect of the medication.

**Illicit Drugs**

Mescaline and lysergic acid diethylamine tartrate (LSD) are hallucinogens that act on specific serotonin (5-hydroxytryptamine: 5HT2) receptors. LSD is active on serotonergic neurons. With these hallucinogens, insight is maintained and there is no evidence of psychosis or delusions. The visual hallucinations may begin as visual distortions, colored patterns, or shapes. They may progress to figures of people and animals, have distortions in size, and involve feelings of fantasy.

Stimulants, such as amphetamine and cocaine, more commonly cause auditory or tactile hallucinations. Cocaine acts on specific serotonin (5HT3) receptors, dopamine receptors, and catecholamine receptors, which are all concentrated in the limbic system. They may begin as simple hallucinations and later develop into vividly colored, complex hallucinations. They are more easily seen with eyes closed or in the dark.19

**Psychiatric Disorders**

Visual hallucinations may be prominent in people with psychoses. However, those with psychosis will also have hallucinations affecting other sensory modalities which differentiates it from Charles Bonnet Syndrome.26 There are many studies that evaluate whether isolated visual hallucinations may be a precursor for developing dementia.14,30,31 In Guerra-Garcia's study (1997), Charles Bonnet Syndrome patients underwent a brain SPECT imaging test.31 SPECT revealed reduced perfusion in the mid-parietal and occipital areas. The hypoperfusion in the parietal area closely corresponds to SPECT patterns that exist in Alzheimer's disease and other forms of dementia. In older adults with Charles Bonnet Syndrome, the presence of other brain disorders should be investigated as a possible cause for early cognitive decline in addition to dementia.

Dementia with Lewy bodies is a common type of progressive dementia. Clinical features of dementia with Lewy bodies include progressive cognitive decline, fluctuations in attention and alertness, recurrent visual hallucinations, and motor features of Parkinson's disease. Symptoms of dementia with Lewy bodies are caused by buildup of Lewy bodies inside brain neurons responsible for memory and motor control.32 Kazui et al. (2009) evaluated glucose metabolism of those patients having dementia with Lewy bodies.25 Results showed glucose metabolism was decreased in the primary visual cortices; however, glucose metabolism was higher in visual association cortices of dementia with Lewy bodies patients who experienced visual hallucinations. The occurrence of visual hallucinations is a result of dysfunction in primary and secondary visual cortices but requires preserved function in visual association cortices.

In a study of patients with AMD, Holroyd and Rabins (1996) found slightly lower cognitive scores for subjects who experienced visual hallucinations compared to cognitive scores of patients without hallucinations were not statistically significant.11 Isolated visual hallucinations were not considered an early symptom of progressive dementia in this study. The relationship between visual hallucinations and development of dementia is still uncertain.

In schizophrenia, visual hallucinations occur more commonly than previously thought. It occurs typically with a delusional component that is not normally seen in organic causes. As schizophrenia progresses, the clarity of hallucinations may deteriorate, which is distinct from Charles Bonnet Syndrome.Table 2 outlines the clinical characteristics which differentiate schizophrenia-related visual hallucinations compared to those originating from organic disease.

| Schizophrenia | Organic disease |
|---|---|
| Waking hours, associated with increased arousal | Nocturnal, associated with drowsiness |
| More paranoia and thought disorder | Less paranoia and thought disorder |
| More personal experiences | Insight |
| Visual and auditory hallucinations | Visual hallucinations |
| Deterioration in clarity of visual hallucinations | Seen in better clarity than remaining vision |
| Delusions | No delusions |

**TABLE 2:** Differentiating schizophrenia-related visual hallucinations versus organic disease-related visual hallucinations

**PATHOPHYSIOLOGY OF CHARLES BONNET SYNDROME**

The true underlying mechanism of visual hallucinations associated with Charles Bonnet Syndrome is not exactly known.1,3,4,17,23,26,33 There are a few well-recognized theories. Hallucinations can result from release by the visual association cortex, acting by loss of cortico-cortical inputs, and alteration of serotonergic processes.19

**Irritative Theory**

According to the irritative theory, diffuse irritative lesions in seizures and migraines26 send abnormal input to the visual association cortices. From there, visual association cortices will generate excitatory discharges transmitted to occipital and temporal lobes,33 which become interpreted as visual hallucinations.

**Release Phenomenon**

The brain is constantly being bombarded with visual stimuli. When people see external visual information, stimuli from visual information will stop activation from within the visual association cortices.17 As a result, no visual hallucinations are released.

According to release phenomenon, any deprivation to the visual system interferes with normal circuitry that inhibits activity in visual association cortex. This lack of inhibition will result in inappropriate excitation of visual association cortices18 and subsequently cause visual hallucinations to be released.This causes visual images to be released from the subconscious to the conscious. In other words, lesions at any level of the visual pathway cause defective electrochemical impulses to be released in addition to the normal impulses. This abnormal visual processing results in visual hallucinations.

Phantom sensations, such as auditory hallucinations, in the recently deaf or phantom tactile sensations in amputees represent similar phenomenon as Charles Bonnet Syndrome except in different sensory domains. Due to the absence of afferent sensory input to the brain, central nervous system increases activity. This increased activity is misinterpreted as hallucinations in the corresponding sensory system.26,54 Some investigators describe Charles Bonnet Syndrome as phantom vision.

Researchers at Harvard-Thorndike General Clinical Research Center conducted a study to determine whether they could induce visual hallucinations by deprivation of the visual system.18 The researchers utilized fMRI to study brain activation in response to stimulation. The results revealed rapid, complete, and prolonged deprivation to the visual system is enough to induce visual hallucinations in fully sighted individuals. It was also found that visual hallucinations occur more frequently with higher levels of visual impairment and bilateral blindness.18 The Release Phenomenon also explains what occurs with sleep deprivation, psychoses, and use of hallucinogenic drugs.

**Deafferentation Theory (Sensory Deprivation Theory)**

Deafferentation theory is the hypothesis that is most widely accepted to explain Charles Bonnet Syndrome. Deafferentation means loss of visual input into the brain will lead to change in excitability of visual association cortex.12 When sensory visual input into cortex is removed from ocular pathology or damage to visual pathways, spontaneous neuronal discharge in visual association cortex occurs, increasing excitability within visual association cortex. As a result, visual hallucinations are released.

The increase in excitability in visual association cortex occurs for a variety of reasons: (1) increase in the number of neurotransmitters released in the presynaptic neuron caused by the presence of increased vesicles; (2) increase in the number of receptors in the postsynaptic membrane caused by prolonged inactivity, which also leads to an increase in the intensity of response; and (3) changes in the amount of gamma-aminobutyric acid and glutamatergic N-methyl-d-aspartic acid within the synapse that has been shown to contribute to hyperexcitability of neurons.45

Because the visual system has plasticity, it can lead to sprouting new axons in damaged areas and reorganization of receptive fields. Therefore, small amounts of remaining stimulus from the retina, optic tract, or other structures within the visual system can elicit visual hallucinations because the neurons are more sensitive.

The sensory deprivation theory is similar to the release theory that abnormal electrochemical impulses are released in the brain. The main difference in sensory deprivation theory is due to the reduction in sensory input to the brain, which causes spontaneous discharge in visual association cortex. This is directly a result of a decrease in visual stimuli caused by ocular pathology.54 In a study, 70% of those with visual hallucinations also suffered from hearing loss. It has been found that people with dual sensory loss in vision and hearing are more likely to suffer from visual hallucinations because of lack of sensory stimulation, thus reinforcing the sensory deprivation theory as cause of Charles Bonnet Syndrome.

**VISUAL PATHWAYS**

After visual information leaves the LGN, it passes along optic radiations and transmitted to the primary visual cortex (Brodmann Area 17). From the primary visual cortex, it will transmit information to the secondary visual cortex (Brodmann Area 18) and finally transfer visual information to higher processing areas of the visual association cortices (Brodmann Areas 19, 37).The primary visual cortex (Brodmann Area 17) is responsible for simple visual hallucinations. The secondary visual cortex (Brodmann Area 18) and visual association cortices (Brodmann Areas 19, 37) are accountable for complex visual hallucinations.

**NEUROBIOLOGY OF VISUAL HALLUCINATIONS**

Comprehending the underlying functional neurobiology will help to understand areas of the visual cortex, which are responsible for stimulating specific visual hallucinations. Neuroimaging studies are providing insight into the science of visual hallucinations. Functional MRI studies have been conducted on patients experiencing visual hallucinations. In these studies, fMRI is used to identify the cerebral location relating to visual hallucinations. Santhouse et al. (2000) found that visual hallucinations were related to increases in activity within specialized areas of the visual association cortex.

Functional MRI studies revealed that when visual hallucinations occur, the occipital cortex has a decreased response to external visual stimuli. The decreased response to sensory input allows endogenous visual stimuli to be perceived as hallucinations.There is a relationship between the content of hallucinations and corresponding regions within the ventral occipital lobe. The ventral occipito-temporal cortex, also known as ventral occipital lobe, is highly specialized for complex visual hallucinations, such as landscapes, figures, faces, people, vehicles, trees, shrubs, and Lilliputian hallucinations.10,13,45,49 The types of visual hallucinations in relation to their specific cortical locations are outlined in Table 3.

| Visual hallucination content | Location |
|---|---|
| Color | Posterior fusiform gyrus |
| Faces | Left middle fusiform gyrus |
| Objects | Right middle fusiform gyrus |
| Visual textures (bricks, fences, maps) | Collateral sulcus |
| Black and white hallucinations | Behind and above the posterior fusiform gyrus |

**TABLE 3:** Type of visual hallucinations related to specific locations within the ventral occipital lobe

**MANAGEMENT STRATEGIES**

In management of these patients, it is important to remember Charles Bonnet Syndrome is a diagnosis of exclusion. Other etiologies that can cause visual hallucinations must be investigated and ruled out before concluding that it is Charles Bonnet Syndrome.1 Charles Bonnet Syndrome may be difficult to discriminate among other comorbidities. Vukicevic and Fitzmaurice created a validated questionnaire to assess characteristics of visual hallucinations.

**Optical Intervention**

The first step in managing these patients with visual hallucinations is to maximize their remaining vision either with spectacles, contact lenses, optical aids, and/or low vision rehabilitation. By improving vision, it is possible to reduce the frequency of visual hallucinations.

In 2004, Eperjesi and Akbarali reported three case studies where visual hallucinations decreased in frequency when patients used optical devices. In these studies, patients were prescribed ground-in prism, monocular telescope, best-corrected spectacles, tinted sunglasses, and nightlights. All of these interventions were effective in reducing or eliminating visual hallucinations for these patients. A hypothesis for the success of these interventions in resolving visual hallucinations was because they provide the sharpest images possible to the retina by reducing blur, glare, and field loss.

**Surgical Intervention**

If surgical intervention is appropriate, treating the underlying cause of visual impairment can halt visual hallucinations. Cataract extraction improved vision and eliminated visual hallucinations in a patient with bilateral cataracts.8 Laser intervention for the treatment of proliferative diabetic retinopathy also reduced visual hallucinations in a patient.8 Laser photocoagulation in a patient with subretinal hemorrhage reduced the occurrence of visual hallucinations.

In a study with 220 patients with neovascular age-related macular degeneration who had treatment with intravitreal ranibizumab injections, 22 of them experienced Charles Bonnet Syndrome. Five of the patients (23%) with Charles Bonnet Syndrome reported an improvement in their symptoms (decrease in frequency, decrease in intensity, or complete resolution of visual hallucinations) if their visual acuity improved after the injection treatment. However, if the patients' visual acuity remained unchanged after the injections, then the visual hallucinations also remained unchanged.

**CONCLUSIONS**

Although awareness of Charles Bonnet Syndrome in the medical literature has increased over the past few decades, there remains a paucity of information about this condition in the optometric and ophthalmologic literature. Criteria for diagnosing Charles Bonnet Syndrome include visual hallucinations, absence of hallucinations in other sensory modalities, partial or full insight that hallucinations are not real, absence of psychological disorders, preserved intellectual functioning, no lower limit of visual acuity, and no specific age.

Knowledge of Charles Bonnet Syndrome allows for proper diagnosis and appropriate management of patients. Recognizing the symptoms of Charles Bonnet Syndrome permits the healthcare professional to share appropriate information about the condition with patients and their families. An increased awareness about Charles Bonnet Syndrome among eye care professionals will benefit and ensure effective management of patients with symptoms of visual hallucinations. If patients experience visual hallucinations, eye care professionals play a critical role in reassuring patients that it can be a normal consequence of their vision loss, informing them and family members about the condition, and coordinating care with other health care professionals to rule out other etiologies of visual hallucinations to effectively manage Charles Bonnet Syndrome. With our patients aging and living longer lives, there will be more individuals affected by vision loss due to age-related conditions. Therefore, we can expect more individuals experiencing Charles Bonnet Syndrome. By recognizing symptoms and identifying signs of Charles Bonnet Syndrome, eye care practitioners will avoid misdiagnosis and/or unnecessary psychiatric or medical treatment causing more distress or burden to patients.

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