Charles Bonnet syndrome is an uncommon condition causing visual hallucination in patients who do not have mental illness.

A pleasant 80-year-old woman who lives independently comes with her daughter to the emergency department complaining of visual hallucinations that have become progressively worse. She does not have a history of dementia or psychosis.

The patient underwent left eye cataract surgery approximately 3 weeks earlier and reports experiencing intermittent visual hallucinations since that time. She described initially seeing family members who were no longer alive. The nature of the hallucinations had become more menacing, however, and she had called the police the evening before this hospital visit. She believed she had seen a man in her house and that he had dropped off a child. She believed the child did not have legs. She was concerned that she was unable to care for the child.

She admitted feeling anxious about the situation but said she had not seen anyone out of the ordinary since she arrived at the hospital. She denied hearing voices and did not exhibit delusional symptoms.

The patient’s past medical/surgical history was significant for hypertension; open angle glaucoma (unmedicated since surgery); recent cataract surgery; clipping for cerebral aneurysm; and dilation for esophageal stricture.

She denied any tobacco, alcohol, or drug abuse. She had no known drug allergies and family history was non-contributory. Her current medications were losartan potassium 50 mg, twice daily; nebivolol 10 mg, twice daily; alprazolam 0.25 mg 3 times daily, as needed for anxiety; and brimonidine eye drops 1 drop and brinzolamide eye drops 1 drop, each 3 times daily. Review of systems was otherwise negative.

On physical examination, she appeared well and in no acute distress. Vital signs were stable. Systemic examination, including neurologic and fundoscopic examination, revealed no gross abnormalities. A mental status examination did not reveal mood or cognitive deficit. She denied any history of underlying mental illness and denied any auditory hallucinations.

Laboratory studies found complete blood cell count and comprehensive metabolic panel within normal limits. No evidence of drug or alcohol use was found. Thyroid-stimulating hormone and vitamin B12 levels also were within normal range. ECG reading was normal. CT of the brain showed postoperative changes from previous fronto-temporal craniotomy for aneurysm clipping. Chronic encephalomalacia was observed in the left frontal and temporal lobes. There was no acute intracranial hemorrhage or cortical infarction seen. Moderate atrophy was observed.

On the basis of the clinical history and examination, a diagnosis of Charles Bonnet syndrome was made. The patient was admitted and examined by Neurology and Ophthalmology. Findings on examinations and evaluations were negative. She denied visual hallucinations during the observation period and resumed her home medications. She was discharged in good condition and directed to follow up with her primary care physician.

Discussion
Charles Bonnet syndrome (CBS) is an uncommon condition that causes visual hallucination in patients who do not have mental illness. The majority of patients with CBS are elderly, with a mean age of 70 to 85 years. Most have some degree of visual impairment or deafferentation of the visual cortex.
Charles Bonnet, a Swiss philosopher, first described the syndrome in 1760 in a publication describing visual hallucination experienced by his grandfather who was blind secondary to cataract but otherwise physically and mentally healthy. It was not until 1937, however, that a Swiss scientist, George de Morsier, labeled the condition as Charles Bonnet syndrome.¹

The diagnostic criteria for CBS remain controversial. The most widely accepted are (1) the presence of formed, complex, persistent or repetitive, stereotyped visual hallucinations in a partially sighted person; (2) full or partial insight into the unreal nature of the perceptions; (3) absence of hallucinations on other sensory modalities; and (4) absence of mental disorders.²

The number of reported cases is increasing as the population ages and the prevalence of ophthalmologic³ and cerebral diseases increases proportionately, including age-related macular degeneration, cataract, glaucoma, diabetic retinopathy, cerebrovascular disorders, and Alzheimer disease. The estimated prevalence is 0.5% to 17%.⁵ It is difficult to interpret, however, because of differences in diagnostic criteria for CBS and in the methods used to evaluate the visual hallucinations.⁶

Visual hallucinations are also under-reported by patients who fear the symptoms represent psychiatric disease or who lack insight into the unreal nature of hallucinations.¹ Up to 60% of patients with CBS are hesitant to tell their physician about their visual hallucinations for fear of being labeled with mental illness or dementia.⁸ A large prospective study in the Netherlands found a history of diminished visual acuity or visual field loss as well as elderly age to be the primary factors correlated with CBS.⁹

Discussion, cont'd
Although the etiology of CBS is unknown, Burk¹⁰ proposes a pathophysiologic model of “neuronal deafferentation.” It is hypothesized that deafferentation caused by retinal or cortical lesions renders neurons more responsive to neurotransmitter release by increasing the number and/or sensitivity of the postsynaptic receptors¹¹ and creating a state of hyperexcitability. Such hyperexcitability occurs in different cortical systems: visual, auditory, vestibular, and motor. Because of the increased sensitivity of the deaffrentated tissue, normal levels of intracortical input may trigger visual hallucination.¹²

Other research has shown that neurosurgical deafferentiation of cortical association areas can generate visual hallucinations.¹³

Visual acuity or visual field loss from any cause that affects the eye, optic nerve, optic chiasm, optic tract, optic radiations, or the visual cortex can cause visual hallucination, or CBS.³

Santhouse et al¹⁴ in 2000 demonstrated through functional MRI (fMRI) how different types of hallucinations correspond to lesions in different areas of the visual cortex. A “map” of hallucinations was drawn in which complex features such as objects and landscapes are related to anterior temporal lobe lesions while activation of the fusiform gyrus produces face hallucinations (fusiform face gyrus). The faces described are cartoon-like, sketch-like, and grotesque, with prominent eyes or teeth. The ventral occipital cortex when activated produces object and scene hallucinations. Eye-centered frames of reference (perseveration images, palinopsia) are related to the parietal lobe.¹⁴

The visual hallucination of CBS may be simple or complex and the characteristic features of these images are not associated with the specific anatomical location of the ocular injury.

The simple hallucinations are also referred to as “elementary” or “non-formed.” They do not include complex imagery. Examples include lights, color, lines, shapes, or geometric designs. Complex, or “formed” hallucinations can include images of people, animals, objects, or a life-like scene.¹⁵

CBS hallucinations occur more often with the eyes open than closed⁴ and patients often describe seeing the hallucinations when they are looking at a white background such as a wall, ceiling, or
piece of paper. The hallucination will often disappear if the patient closes his eyes or looks away. The duration of the hallucination ranges from less than 1 minute to continuous. Most patients report duration of several minutes. The frequency is variable; the majority of patients experience hallucinations multiple times a day or a week. Associated symptoms depend on the underlying disorder producing the visual loss.

The strongest risk factors for CBS are bilateral visual system impairment, declining visual acuity, cerebral damage, cognitive defects, social isolation, and sensory deprivation.

The following are some other closely related diagnosis that should be considered in patients with visual hallucinations:

**Peduncular hallucinations.** Peduncular hallucination is a rare manifestation of lesions (usually stroke or neoplasm) affecting the midbrain, in particular, the paramedian reticular formation. Similar syndromes have been described with pontine and thalamic lesions. The hallucinations are complex and binocular, involving both visual fields. The content of the complex imagery varies and usually described as vivid and colorful. The hallucinations may have associated tactile and auditory content.

**Migraine.** Approximately 15% of individuals with migraine experience aura. Visual aura occurs in 90% of people with migraine aura. Migranous visual hallucinations are usually simple and typical linear or geometric (eg, zig zag lines) in appearance. The usual duration for migraine aura is between 4 and 60 minutes. Common associated symptoms include headache and other feature of migraine: nausea, vomiting, photophobia, and phonophobia.

**Seizure.** Visual hallucination of epileptic origin can be simple or complex. Epileptic hallucinations are usually brief—only a few seconds—unless the seizures are continuous. For patients in whom seizures are suspected, a complete neurologic evaluation and electroencephalogram (EEG) should be performed.

**Neurodegenerative disease.** Visual hallucinations are a core clinical feature of dementia with Lewy bodies (DLB) and are also common in Parkinson disease (PD). In DLB and PD, the hallucinations are characteristically complex, binocular, and occur throughout the entire visual field. Descriptions range from well-formed images of people or animals, to more abstract visions such as shapes or color. In DLB, visual hallucinations occur in approximately two-third of patients and occur early in disease course. Parkinsonism, visual hallucinations, and prominent fluctuations of cognition or alertness are characteristic of DLB. In PD, visual hallucinations are more prevalent later in the disease course. Dopaminergic medications can contribute substantively to their occurrence, as well as to other manifestation of psychosis in patients with PD. Visual hallucinations are relatively uncommon in Alzheimer disease, particularly early in the disease course. When present, these often reflect a superimposed delirium, medication effect, or vision loss.

**Alcohol and drug use.** Alcohol and benzodiazepine withdrawal often produce complex hallucinations with vivid imagery. These are often continuous and are associated with agitation, tremulousness, and autonomic hyperactivity.

**Psychiatric illness.** Most visual hallucinations in psychiatric illness are complex. Auditory hallucinations are approximately twice as common and typically accompany the visual hallucinations. The content of the hallucinations is usually disturbing and antagonistic, and symptoms of depression, mania, anxiety, disordered thought, or delusions are usually present.

**Metabolic encephalopathy.** Patients with metabolic encephalopathy are typically confused, often agitated, with other psychotic features, including auditory or tactile hallucinations and delusions. Etiologies are often multiple can include CNS or systemic infection; hypoxia; medications; and metabolic disturbances, such as hepatic or renal failure, electrolyte imbalance, and hypothyroidism.

**Treatment of Charles Bonnet Syndrome**
Treatment is individualized according to the degree the patient is disturbed by their symptoms. Sometimes reassurance may be all that is required. Patients can also be taught to suppress the
hallucinations by closing their eyes or looking away. Increasing arousal and visual stimuli (eg, increased illumination) and reducing social isolation are also reported to be helpful for individual patients, but these techniques have not been systematically studied. Patients who experience continuous hallucinations or disturbing imagery may require pharmacologic treatment with a newer (atypical) antipsychotic agent. Olanzapine and quetiapine have shown benefit without significant side effects. Carbamazepine, alone or in combination with clonazepam, also has been reported to be successful in treating CBS hallucination.

Take-Home Points

- Hallucinations are perceptions in the absence of an external stimulus. When visual hallucinations follow marked visual acuity loss, in the absence of cognitive impairment, the condition is termed "Charles Bonnet syndrome" (CBS).
- The hallucinations of CBS may be simple or complex. These typically last a few minutes and usually recur frequently, daily or weekly.
- CBS can be attributed to “neuronal deafferentiation.” Therefore, lesions should be sought not only in retina, but also in the occipital lobe, visual cortex, and related association area.
- Reassuring the patient is important and may be all that is required. Patients may be able to temporarily suppress hallucinations by closing their eyes or looking away. If hallucinations are troubling to patients, a trial of low-dose atypical antipsychotics, cholinesterase inhibitors, or antiepileptic drugs is suggested.

References


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