

# Neuro-optometry Seminar: Cortical Blindness and Charles Bonnet Syndrome

## Introduction to Neuro-optometry Seminar:

This course will highlight two different conditions, cortical blindness and Charles Bonnet Syndrome. Both are commonly found in low vision practices and hospital-based settings. The purpose is to make eye care practitioners aware of these conditions and allow them to make confident diagnoses and proper referrals.

## Cortical Blindness

### Introduction:

Cortical blindness is characterized by impaired vision resulting from an insult to the visual cortex. Patients with cortical blindness will often present with healthy ocular structures and normal pupils, but best-corrected vision will be count fingers or worse. Cortical blindness should be suspected in cases of sudden unexplained vision loss, especially in older patients who are at risk for stroke.



Image highlighting the location of the visual cortex, seen in yellow.

### Case Presentation:

A 71 year-old inpatient Hispanic male was referred to the Eye Clinic for a low vision examination. He had recently been seen by the Ophthalmology Clinic for a comprehensive eye exam. He presented in a wheelchair and was unable to transfer given his ataxia and low vision. The patient also presented with mild binocular mucous discharge, which had begun just yesterday morning. He had been placed on

erythromycin ointment qhs and artificial tears qid by his primary care provider since the onset of symptoms, with minimal improvement overnight in amount of discharge.

The patient's main goal was to be able to see gross shapes on the television screen. Being illiterate, he had no goals for reading or computer work. As an inpatient, he had his food organized into compartments for him, and he was able to feed himself without problems. His ocular history was significant for cortical blindness diagnosed a year ago. Prior to that, he had an absolute left hemianopsia and right quadrantanopsia, but was still able to ambulate using a white cane. His medical history was significant for hypertension, diabetes with neuropathy, mixed hyperlipidemia, ataxia, and a history of multiple strokes, with the most recent stroke being 4 months prior to this examination. Findings from a recent brain MRI demonstrated "extensive post-ischemic encephalomalacia of the bilateral cerebellar hemispheres and the bilateral occipital lobes, more extensive in the right occipital lobe." Medications included Clopidogrel, Insulin, Metformin, Simvastatin, Benzepiril, and Haloperidol.

Best-corrected visual acuities were HM at 1ft in the right eye, and HM at 3ft in the left eye. Pupils were equal, round, and reactive to light, with no afferent pupillary defect. Extraocular muscles were smooth, accurate, full and extensive OU. Refraction revealed no improvement in vision with +/- 6.00D changes. A yellow tint fit-over helped improve contrast. Anterior segment was remarkable for trace superficial punctate keratitis in both eyes, as well as a central linear stromal scar OD and a small round peripheral rust ring scar OS, and 3+ nuclear sclerotic cataracts OU. Examination of the posterior segment revealed clear media in both eyes, with mild temporal pallor in both optic nerve heads and cup-to-disc ratios of 0.40 in each eye. Mild retinal pigment epithelium mottling within both maculae was also present.

The patient was provided with a pair of yellow tinted glasses and referred to the Blind Rehabilitation Outpatient Services for demonstration of additional assistive devices and orientation and mobility training. He is being followed yearly by ophthalmology for ocular health checks.

#### **Discussion:**

Sudden bilateral cortical blindness with normal pupillary responses was first described in 1920 by Meyer. He postulated that an occipital lobe infarction followed by resulting vision loss was caused by focal compression of the posterior cerebral artery (PCA). Years later, the development of computed tomography (CT) and other imaging techniques would confirm his hypothesis. In 1977, Nosborn demonstrated the first CT finding of an occipital lobe infarct.

According to the Los Angeles Latino Eye Study (LALES), of all the individuals examined who were legally blind, 5% of individuals were blind due to cortical blindness. Cortical blindness can present in both transient and permanent forms.

#### **Signs/Symptoms:**

Symptoms are characterized by bilateral severe or complete loss of vision. The ability to perceive motion and light usually is preserved (Riddoch phenomenon), but static objects remain

undetected. Vision loss is usually sudden, but may be the result of successive strokes that continually limit the patient's visual field, as with the case discussed earlier.

A patient will present with count fingers (CF) or hand motion (HM) vision OD and OS. Cases of no light perception (NLP) vision have been reported but are extremely rare. Visual fields will also be constricted in both eyes. Pupils will be normal, anterior segment should be grossly unremarkable, and the patient should have overall normal fundi and optic nerves. Even if visually critical ocular structures are compromised, the degree of poor acuities will still be unjustified by the amount of ocular damage observed.



In rare cases, individuals will deny their blindness, in what is called Anton Syndrome (or Anton-Babinski Syndrome). Patients will be alert, cooperative, and communicative, but will be unable to perform routine work. Case reports on Anton Syndrome describe individuals as frequently running into objects and having minor accidents while ambulating, though they will not admit to their blindness. The denial of blindness is explained by confabulation, a type of memory disturbance seen commonly with brain damage and dementia. In confabulation, one fabricates distorted memories without an intention to deceive others, which is different from malingering because the patients are unaware the information is false. According to Scharli et al, Anton Syndrome occurs in 1 in 9 patients with bilateral vision loss.

Visual hallucinations may also be present with cortical blindness, in what is called Charles Bonnet Syndrome (CBS). With Charles Bonnet Syndrome, patients are aware that they are blind and know that the hallucination is false. See the other half of this course for full details on CBS.

### **Pathogenesis:**

Cortical blindness is caused by focal ischemia or damage to occipital lobes resulting from disruption of one or both posterior cerebral arteries. This consequently damages the geniculocalcarine pathway (aka optic radiations), causing disruption to V1 and loss of vision. One should keep in mind that because the occipital cortex has large macular representation, macular sparing (small island) may be present and complete vision loss may not occur.

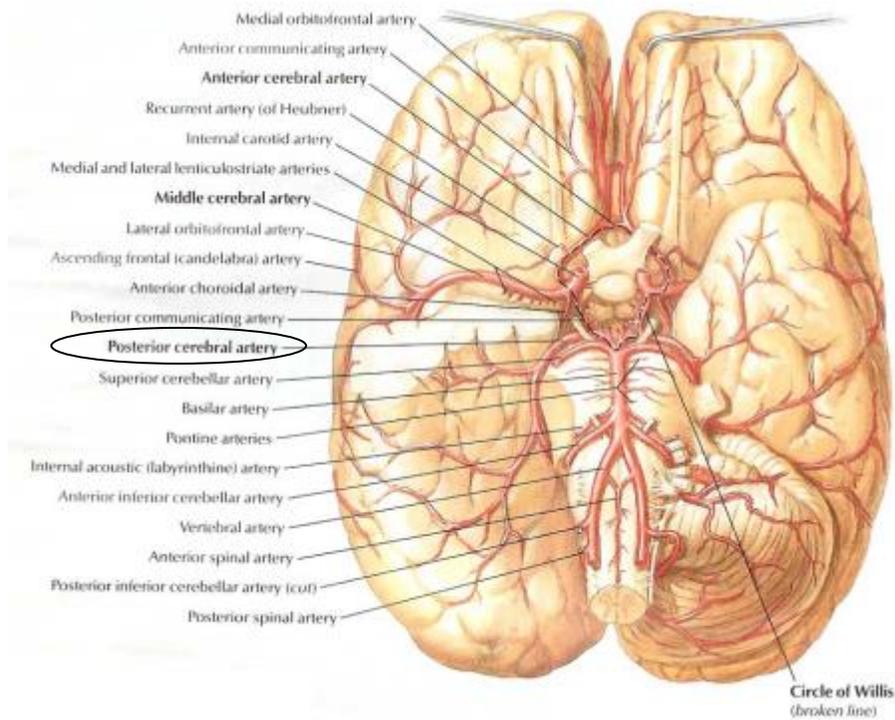


Image of vascular circulation in the brain, the location of the posterior cerebral artery is highlighted here.

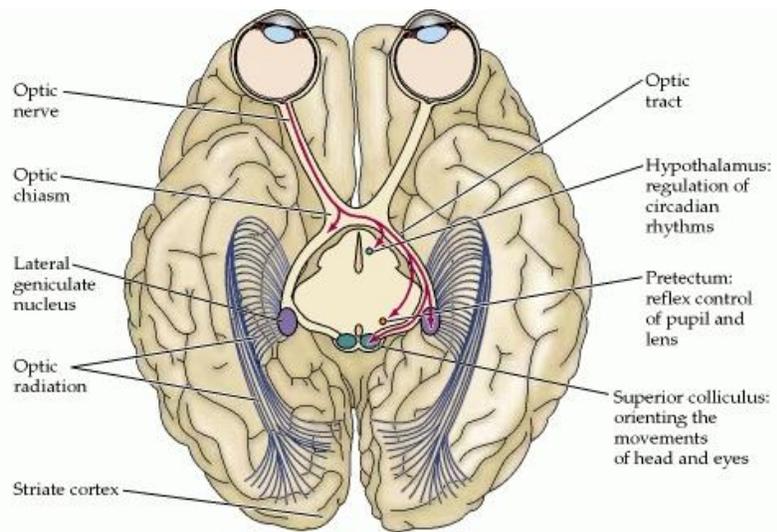


Image of visual pathway.

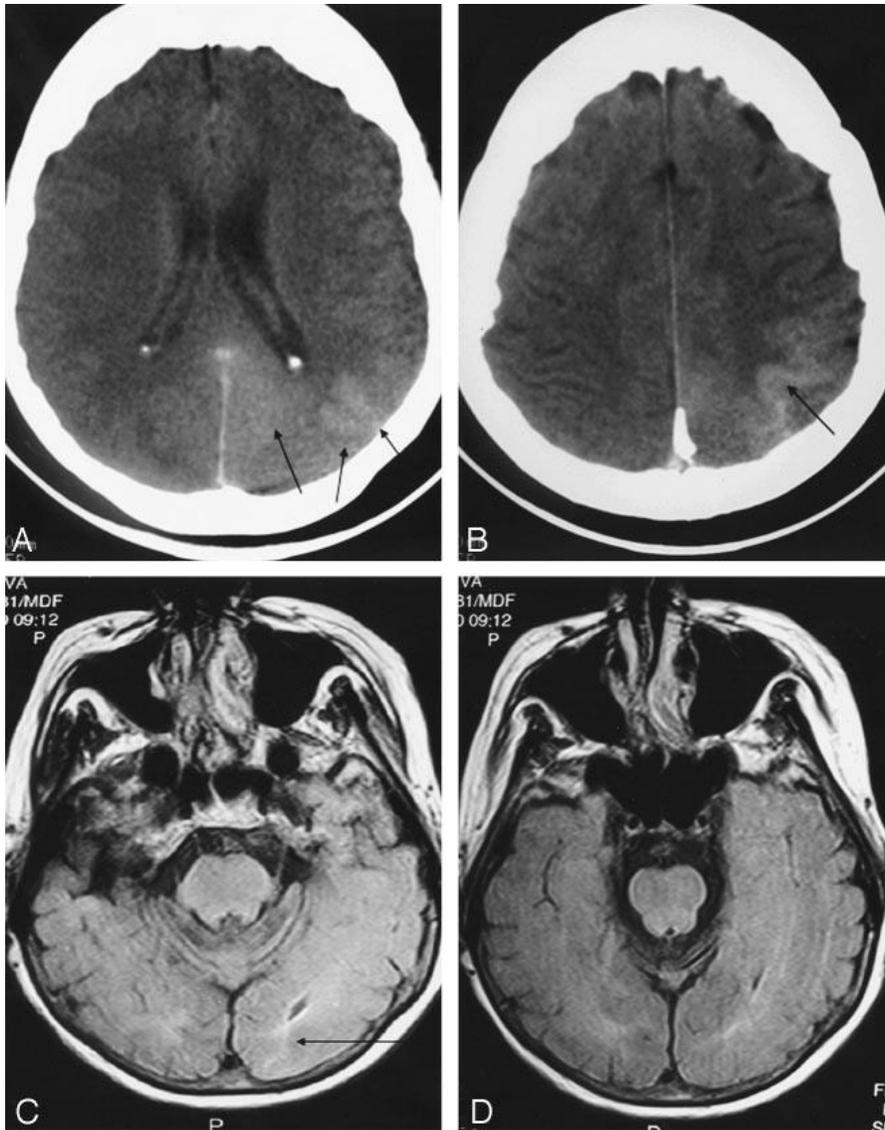
## **Etiology:**

There are multiple etiologies for cortical blindness. When describing profound vision loss in children, the preferred term is Cortical Visual Impairment (CVI). CVI may present congenitally, as a result of traumatic brain injury (TBI) due to shaken baby syndrome or accidental head injury. Encephalitis, meningitis, and ischemic perinatal stroke are also known congenital etiologies for CVI. Because it is near impossible to assess a patient's vision at such an early age, one must look for abnormal visual responses such as brief fixations or blunted social gaze. Looking for cues such as poor hand-eye coordination can help. In addition, these patients will often demonstrate other problems associated with brain damage, such as cerebral palsy or intellectual disabilities.

Stroke is the most common cause of acquired cortical blindness, comprising of around 35% of cases. Vision loss is usually sudden and very severe. While acute infarcts will demonstrate hyperintensity signals on CT or MRI, these areas of the brain will darken over time as encephalomalacia, a process by which brain material is replaced by water, occurs.

Cardiac surgery, especially a Coronary Artery Bypass Graft (CABG) procedure, is the second most common cause of acquired blindness, comprising of around 20% of cases. Fortunately most individuals will recover their vision, but full recovery may take up to months. The exact pathogenesis of vision loss is unknown, though multiple mechanisms have been suggested to explain the cerebral dysfunction. It is possible that the brain may become anoxic from hypoperfusion during surgery. Ischemic neuronal damage to the calcarine cortex is common in individuals who do not survive cardiac surgery. Other etiologies may involve focal cerebral hemorrhages, or patchy ischemia due to a blood, fat, or air embolism. Damage to the brain secondary to vasospasm has also been suggested.

Cerebral angiography comprises around 12% of cases of cortical blindness, and nearly all cases are transient. The onset of blindness is usually rapid (within minutes) of the procedure, but can occur up to 12 hours after the procedure. By 12 hours of onset, the vision should demonstrate significant recovery, although full recovery could take days. The incidence of cortical blindness is different depending on type of contrast agent. For nonionic agents, there is a 0.3-1% chance of vision loss. This number rises to 4% when iodinated contrast agents are utilized. It is postulated that damage to the blood-brain barrier along with a direct neurotoxic effect of the contrast agent may be responsible for the vision loss. CT scans will usually demonstrate bilateral leakage of contrast in the occipital cortices. Please see Figure 1 and Figure 2.

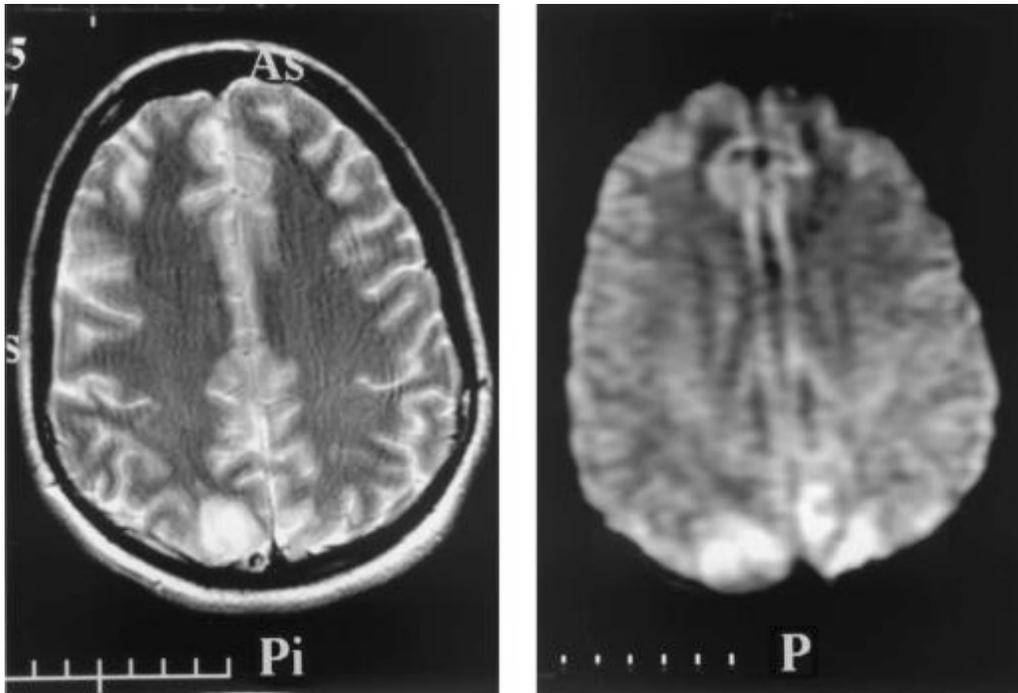


*Figure 1. 74-year-old woman who developed cortical blindness 1 hour after cerebral angiography. Axial noncontrast CT reveals hyperattenuation in the left parietooccipital cortex (A,B) without mass effect. FLAIR MR images (C,D) show hyperintensity in left occipital cortex (long black arrow).*



*Figure 2. FLAIR MRI of 45 year old woman who developed cortical blindness 12 hrs after CT scan. She was disoriented to time and place, and developed Anton Syndrome. The small black arrows show contrast uptake in bilateral occipital cortices.*

Pre-eclampsia is another common cause of transient cortical blindness, occurring in up to 15% of patients with this condition. The reduction in vision can occur in several hours to days of the blood pressure elevation, and vision loss has been reported to last from 4 hours to 8 days. Radiologic findings will reveal petechial hemorrhages with focal areas of edema. While the exact pathogenesis is unknown, there are a few mechanisms whereby normal cerebral autoregulation can be overcome by severe hypertension. Within the brain, there appears to be a disparity in cerebral regional blood flow, with the anterior circulation being receiving more innervation than posterior circulation. Therefore, any insult would appear to have a preferential predilection of symptoms focused within the posterior circulation. One theory suggests that with vasospasm there is consequent ischemic injury and cytotoxic edema that damages the occipital cortex. Another theory suggests that increased capillary permeability and resulting vasogenic edema are responsible for vision loss. Please see Figure 3.



*Figure 3: During a pre-eclamptic episode that forced an emergency Cesarean section, T2 weighted (left) and diffusion weighted (right) imaging revealed high intensity signals in bilateral occipital cortices. After delivering the baby, the patient reported light perception vision in both eyes.*

Other acquired forms of congenital blindness are traumatic brain injury (TBI) to the occipital lobe, and more rarely, neoplasm in the brain as a result of a meningioma or metastasis.

**Risk factors:**

For transient forms of cortical blindness, risk factors include pregnancy and imaging procedures with contrast. It is estimated that up to 8% of individuals in the United States develop pre-eclampsia during pregnancy, with the percentage being even higher in undeveloped countries. There appears to be a higher incidence in Black and Hispanic women over Whites.

For permanent cortical blindness, stroke risk factors, particularly the modifiable ones, need to be addressed with the patient's primary care provider. Without proper management unilateral occipital strokes can progress to bilateral strokes. Please see Table 1.

Uncontrollable Risk Factors	Controllable Medical Risk Factors	Controllable Lifestyle Risk Factors
Gender (M>F)	High Blood Pressure	Tobacco Use and Smoking
Age (older>55)	Atrial Fibrillation	Alcohol Use
Race (African American, Hispanic, Asian American)	High Cholesterol	Physical Inactivity
Family history of vascular disease	Diabetes	Obesity
Prior history of stroke	Atherosclerosis	
	Circulation Problems	

Table 1  
the

lists out

uncontrollable risk factors, controllable medical risk factors and controllable lifestyle risk factors for cortical blindness.

**Ocular work-up:**

In working up suspected cases of cortical blindness, testing visual acuity at distance can provide more information than checking acuity close-up when the patient still has a small island of vision. Given the fact that strokes are the primary cause of cortical blindness, an easy test to perform in office is to check blood pressure. One should be well familiar with TIA/stroke-like symptoms and inquire about them accordingly. It is important to remember that multiple strokes can occur, and recent absence or presence of symptoms should be elicited at each visit. Functional vision loss must also be ruled out, which may include using the nystagmus drum or tangent screen visual fields.

**Management:**

Ocular management involves low vision referral and orientation/mobility training. In most cases the patient will have had a CT scan and/or MRI of the brain in the emergency department, but ordering these tests may be prudent in cases of newly worsened acuity. Further management should involve notifying the patient’s primary care provider (PCP) of the findings and placing a consult with neurology or stroke clinic to have stroke risk factors addressed.



### **Prognosis:**

Visual prognosis will depend on etiology. As expected, severe bilateral occipital lobe infarcts have the worst prognosis. Blindness is permanent in 25% of these patients. Knowing the specific location of damage can be helpful. A study by Cioni et al. revealed that damage to the optic radiations resulted in poorer visual function than injury to the visual cortex, and overall improvement was noted more frequently (78%) in patients with cortical damage than in those with periventricular white matter injury (22%). Patients who are under the age of 40, have no history of hypertension or diabetes mellitus, and do not have associated cognitive, language, or memory impairments tend to have better final visual acuity.

### **Conclusion:**

In examining cases of cortical blindness, it is important to look at the systemic risk factors which may predispose the patient to stroke. It is also important to inquire about recent surgeries and imaging. As cortical blindness is only a manifestation of a deeper problem, involving other medical providers in your care of the patient is an obligation we have as eye care providers.

## **Charles Bonnet Syndrome (CBS)**

### **Introduction**

Charles Bonnet Syndrome (CBS) is defined as the occurrence of visual hallucinations in intact cerebral functioning patients, secondary to visual deprivation. It is often due to some form of ocular pathology. CBS-induced visual hallucinations are considered “pseudohallucinations”, which refers to a sensory experience occurring without external stimulation of the eyes. The patient is fully aware of the unreality of the sensory experience.

CBS usually occurs in the visually impaired, elderly population, with a mean age of 78.2 years old. There have been case reports of children sustaining rapid visual loss that have developed CBS as well. Because the elderly population is at most risk for ocular pathology and visual deprivation, CBS is also more prevalent in this population. While the prevalence of CBS is around 11-15% of low vision patients, these

numbers likely underestimate the true number of cases because these individuals are often misdiagnosed with dementia or psychosis. Also, patients are often reluctant to report symptoms with the stigma of a dementia or psychosis diagnosis. Studies differ on whether there is a gender bias.



## Case Report

An 89 year old Caucasian male, CW, presented for a comprehensive eye exam. CW voluntarily reported often seeing people or images that are not really there. The patient reported that the onset was very gradual and began within the last couple months, and that the images or people never speak with him. He is fully aware that the images or people are not real, and is not bothered or afraid of the hallucinations. He proceeded to share a story of a recent hallucinatory experience: "I went to the bank recently and I thought I saw a million dollars on the ground. I walked over and tried to pick it up, but when I touched it, it disappeared and I realized it was not really there." The patient had no other visual complaints and denied changes in vision since his last eye exam one year ago.

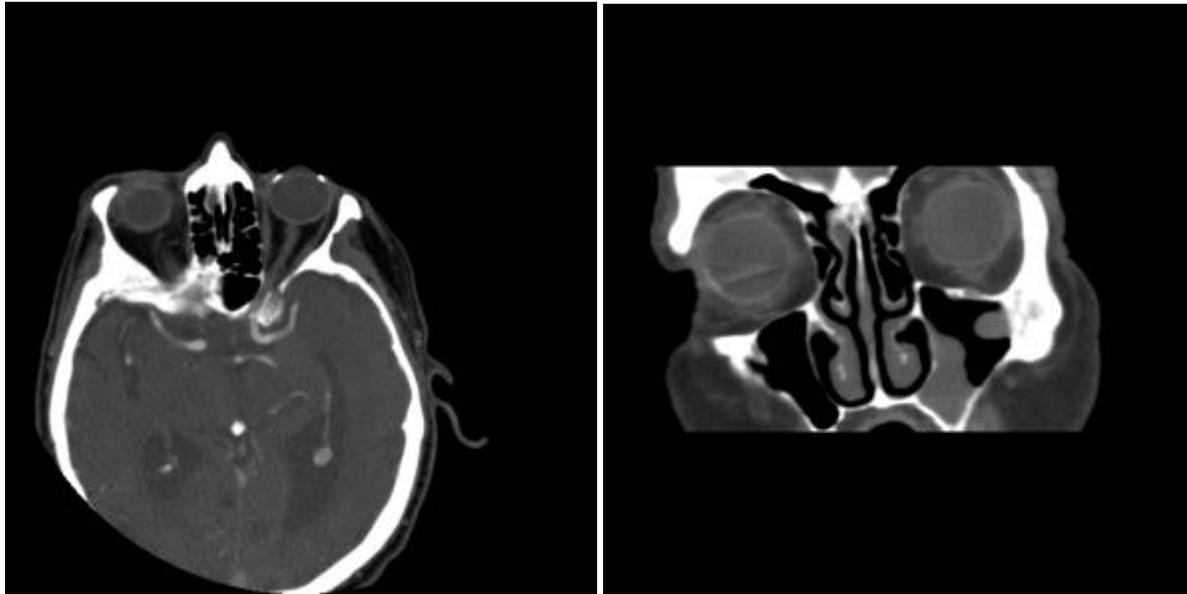
The patient's ocular history was significant for narrow angle glaucoma OU status post bleb and laser peripheral iridotomy (LPI), age related dry macular degeneration AREDS category III OU, corneal haze secondary to surgery OU, and pseudophakia OU. Significant ocular medications included Timolol qAM OS and OcuVite with lutein PO BID.

Medical history was significant for hypertension and acne rosacea. Recent vitals revealed mildly reduced platelet levels, but was otherwise unremarkable. Recent blood pressure was unremarkable at 123/67.

Best-corrected visual acuities were 20/400 OD and 20/150 OS. Extraocular muscles were smooth, accurate, full and extensive. Confrontational visual fields were full to finger counting OD and OS. Pupils were surgically non-reactive and fixed OD and minimally reactive OS, with no afferent pupillary defect. Slit lamp exam was significant for 2+ diffuse corneal haze OD>OS and Descemet's folds OD>OS. In addition, there were 2-3 dense areas of stromal opacity OS, each smaller than 1/2 mm in diameter. Throughout the course of the exam, the patient repeatedly described seeing flowers, smiley faces, and little animals. A dilated fundus exam revealed hazy media OU, with a moderate-large cup to disc ratio consistent with glaucoma OU. Macular findings included extensive drusen, retinal pigmented

epithelium mottling and non-central geographic atrophy OU consistent with macular degeneration AREDS category III. A spectral domain OCT of macula confirmed the absence of choroidal neovascularization OU.

Final assessment included all previous diagnoses and visual hallucinations consistent with Charles Bonnet Syndrome. In order to rule out visual pathway and other central lesions known to cause visual hallucinations, a CT without contrast of the orbits was ordered. The CT scan revealed no intraorbital masses or other visible abnormalities. The visualized portions of the optic pathways were unremarkable in appearance. Image below shows CW's CT scans.



## Discussion

### *History:*

CBS is named after Charles Bonnet, a renowned Swiss naturalist, philosopher and biologist. He was the first to describe these hallucinatory experiences in his own grandfather, Charles Lullin in 1769. Lullin had lost his vision to bilateral cataracts. He had intact cognitive function and was fully aware that the hallucinations were not real. He described silent visions of men, women, birds, carriages and buildings, varying in size, shape, and place. Ironically, due to unknown etiology, Charles Bonnet developed some vision loss in the later decades of his life and himself developed CBS as well.



Image of Charles Bonnet (1720-1793).

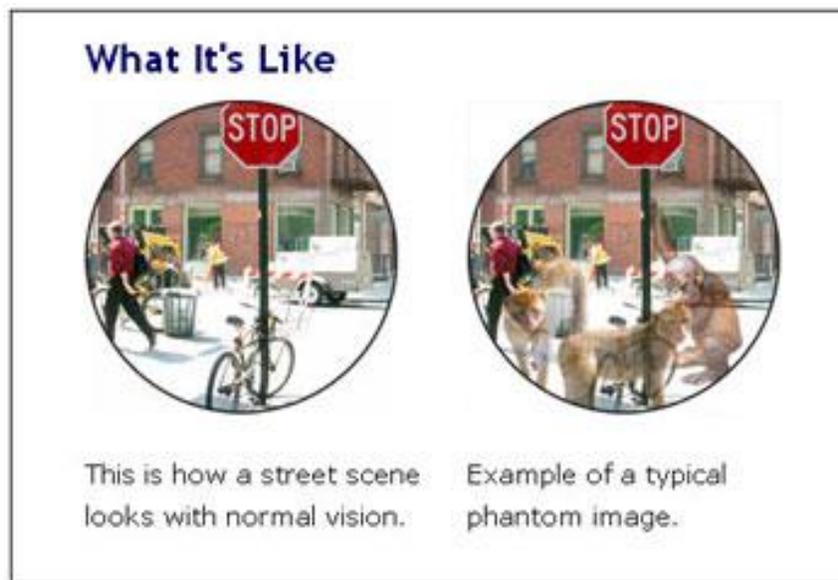
***Features:***

Patients that experience CBS have been reported seeing persons, faces (regular or distorted), animals, figures, shapes, and earlier stages of themselves. The images are always projected in external space and are very organized, defined and vivid. Patients with CBS often have profound vision loss, so real life images are blurry in comparison to the hallucinations. The stark contrast between blurry real life objects and clear hallucinatory objects allows patients to distinguish hallucinations from real life objects. Hallucinations can also be in black and white or color, and can change from initially being very simple into developing into a complex hallucination or vice versa. They can be static, dynamic, or moving “en bloc,” referring to an absence of movement from the image itself but the image moves relative to the space around it.

The hallucinations can last for seconds to minutes to hours. Triggers include sensory reduction, fatigue, stress, and low or bright illumination. Alleviating factors include executing ocular saccades, looking directly at the image, approaching the image, or conversing with the image. Sometimes the image can spontaneously disappear on its own without any external alleviating factors. Closing the eyes has been reported to act as both a trigger and an alleviating factor. Most hallucinations are exclusively visual, unaccompanied by noise or other senses. However, there are few case reports of individuals with CBS reporting auditory hallucinations alongside the visual.



An interpretation of CBS pseudohallucination: the hallucination image of the man in the sombrero hat is more vivid and clearer compared to the surrounding blurry image of the real world. There is also an absolute central scotoma present here.



Another interpretation of CBS pseudohallucination.

There are three time phase categories of CBS: episodic, periodic, and continuous. Episodic CBS, the rarest of the three, refers to episodes of CBS occurring and lasting from days to months and then permanently resolving. Periodic CBS refers to having phases of hallucinations and phases or remissions from these hallucinations. Continuous CBS refers to ongoing hallucinations without hallucination-free intervals. Periodic and continuous are far more common.

Patients have very different reactions to developing CBS. Depending on the hallucination, patients can be indifferent, curious, irritated or terrified. Most patients are fearful of being ostracized by others or labeled as psychiatrically unstable or demented.

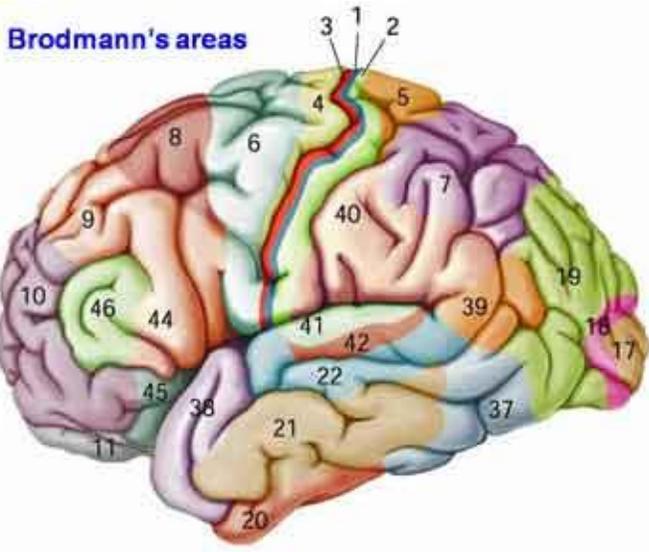
Despite the variations in sensory experience, the patient should always be aware of the unreality of the hallucinations. While insight into this fact is not always immediate and can sometimes fluctuate, it is critical that there is at least a small component of acknowledgment of the unreality of the hallucinations. Patients with CBS have been reported with variable mental states, but this area remains controversial because most definitions of CBS agree that there should be an absence of depression or other psychopathology. Some studies state that visual hallucinations are early markers for dementia. Patients that acquire Charles Bonnet Syndrome with the addition of some mild form of cognitive impairment can be further labeled as "The Charles Bonnet Plus Syndrome." This diagnosis is rare and clinicians often find it unnecessary to subcategorize to this level.

### ***Risk Factors***

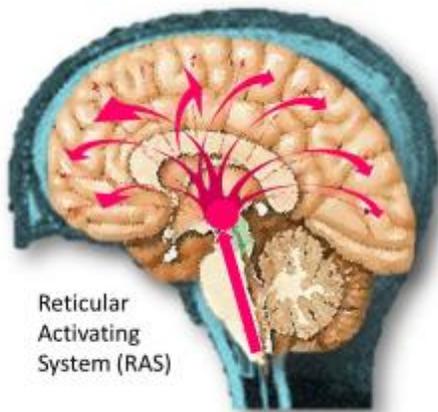
The biggest associated risk factor for developing CBS is acquired visual impairment. Most CBS cases have observed vision loss of 20/60 or worse and present more commonly with bilateral vision loss. Studies that support the association of CBS and vision loss highlight cases where bilateral age related cataracts temporarily cause visual impairment in elderly patients. Once cataract extraction is performed, vision immediately returns to normal and the hallucinations either improve or cease all together. However, some studies argue against the association of CBS and vision loss. These studies cite cases where CBS gradually ceases even with further progression and deterioration of vision. Other associated risk factors include social isolation, shyness, cerebrovascular disease, fatigue, disturbances of vigilance, stress and suggestibility. Ultimately, any type of sensory deprivation, more likely in visual form, can trigger the development of CBS.

### ***Neuroanatomy***

The neuroanatomic basis of complex visual hallucinations is localized to Brodmann's areas 18 and 19 with elementary sensations originating from area 17. Activation of different cortical areas seems to produce different images. In addition, functioning magnetic resonance imaging (fMRI) during hallucinations show a decrease in response to exogenous visual stimulation in conjunction with disinhibiting endogenous visual memories. The combination of these events helps spawn hallucinations. Moreover, there are studies that find a strong association between drowsiness and hallucinations. In particular, the reticular activating system (RAS), which plays a part in alertness and suppressing hallucinatory tendencies, is dampened when drowsy. It is thought that the dampened signals of RAS gives rise to hallucinations.



This image shows the location of area 17, 18, and 19.



This image shows the location of RAS.

***Diagnostic criteria***

There are no standard diagnostic criteria for CBS; however, many have been proposed. Two which are more widely accepted are suggested by Podoll et al and Gold/Rabins. Podoll et al. suggests three conditions for diagnosis. First, the predominant symptom is visual hallucinations in elderly individuals with specifically normal mental health. Second, no delirium, dementia, negative impact on intellectual capacity, and cognitive deterioration can be present. Examples may include the affective syndromes, paranoid developments, psychosis, intoxication or neurological disease. Third, loss of vision must be present, which may or may not be a consequence of ocular disease. Gold/Rabins also had three conditions for the diagnosis of CBS. First, visual hallucinations must be formed, complex,

persistent/repetitive and stereotyped. Second, insight to these hallucinations must be fully or partially retained. Third, hallucinations in other modalities or senses must be absent. These conditions may provide guidelines for diagnosing CBS, but do not serve as a definitive diagnostic criteria.

### ***Associated conditions***

Anything causing an acquired deprivation of visual senses can be associated with CBS. A few conditions that have been reported include: age related macular degeneration, cataract, corneal opacities, choroideremia, glaucoma, retinal detachment, enucleation, multiple sclerosis with optic neuritis, retinitis pigmentosa, occipital lobe infarction with both homonymous hemianopia, bilateral loss of vision, venous congestion of occipital cortex due to AV malformation, vertebra-basilar insufficiency, grief reaction, and cortical blindness. See the other half of this course for full details on cortical blindness.

### ***Theories of pathogenesis***

There are many proposed theories that attempt to explain CBS. A few of the more widely accepted theories include: *sensory deprivation-phantom vision, dreams and hallucinations, social isolation, psychological factors, and anomalies of cerebral perfusion.*

The *sensory deprivation-phantom vision theory* suggests that hallucinations are memories of previous perceptions. In normal sighted individuals, these memories are suppressed because there is a constant influx of visual stimulation. However, in sensory deprived individuals, the visual cortex is not receiving the normal afferent input it is accustomed to. Therefore, to fill the void, memories of previous perceptions begin to surface into a person's consciousness. These memories turn into hallucinations because there is no longer active visual stimulation to suppress them. Support for this theory lies in cases of CBS where hallucinations disappear after normal or excessive visual stimulation is restored, as in the cases of bilateral cataract extraction.

The *dreams and hallucinations theory* parallels normally sighted individuals who are asleep to alert and awake CBS individuals. This theory suggests that there is always a stable and constant threshold of sensory input during sleeping and waking hours. Throughout the day, a person's daily tasks involves being bombarded with a variety of stimulation. Therefore, in the waking hours of a normally sighted individual, sensory input remains above threshold. However, during sleep, sensory input diminishes and drops below threshold. As a result, the cerebral cortex becomes super sensitive to background activity giving rise to hallucinations or "dreams." Patients with CBS develop hallucinations because even during waking hours, their visual sensory input remains fixed below threshold. As a result, they are able to hallucinate or "dream" just as a normal sighted person can do only while asleep.

The *theory of social isolation* suggests that an individual subjected to social isolation is in a general state of sensory deprivation. When this occurs, hallucinations develop to fill in the void. A sensory deprivation study conducted by Heron et al obtained findings to support this theory. The experiment used the researchers themselves as test subjects. Each person was subjected to one full day of sensory deprivation in an isolated room. All test subjects invariably experienced some form of visual hallucinatory experiences by the end of the day. The hallucinations were described as being first

elementary, then complex with movement. In addition, Heron et al found that hallucinations had a tendency to increase during periods of reduced alertness. This further supports alertness playing a critical role in the likelihood of developing hallucinatory experiences.

The *theory of psychological factors* suggests that a psychological desire to see under partial vision loss drives hallucinations to develop. There is not much research in this area to support this theory.

The *theory of anomalies of cerebral perfusion* suggests that any type of cerebral non-perfusion or anomaly can cause hallucinations to occur. A few examples include vertebro-basilar insufficiency with ischemia or visual pathway, asymmetrical hyperperfusion in the lateral temporal cortex, corpus striatum and thalamus, reduced occipital perfusion, and mid-parietal and occipital hypoperfusion. Anton Syndrome represents an extreme, where patients deny their blindness and convince themselves that they are still visually functioning individuals. For more details on Anton Syndrome, please see the Cortical Blindness section.

### ***Differential Diagnosis***

Since CBS is a diagnosis of exclusion, various differentials for hallucinatory causes must be ruled out before CBS can be formally diagnosed. Differential diagnoses for CBS include any neuropsychiatric condition, emotional disturbance and decreased alertness. Some examples include dementia, lewy body dementia, delirium, hypnagogic hallucinations, Alzheimer's dementia, Parkinson's disease, peduncular hallucinosis, levodopa-induced hallucinations, migraine coma, schizophrenia, medication and epilepsy.

Dementia, lewybody dementia or delirium can present like CBS, but unlike CBS, patients will have impaired attention, disorganized thought processes, and abnormalities of sleep or orientation. Hypnagogic hallucinations can occur immediately before sleep usually while the eyes are closed and individuals are transitioning between that delicate state of being awake and asleep. This phenomenon occurs in about 1/3 of normal patients, and sometimes the visual hallucinations can be accompanied by auditory sensations. Parkinson's disease is characterized by impaired visual function, clear sensorium, absence of hallucinations of other modalities, impaired cognition, and depression. These symptoms can be dependent on dose or duration of anti-Parkinsonian medication or duration of illness. Other differentials like levodopa-induced hallucinations, migraine coma, medication, Alzheimer's disease, strokes, serious mental illness (i.e. schizophrenia) and other brain conditions (i.e. peduncular hallucinosis) can affect the visual pathway and visual cortex involved with vision. These conditions can cause hallucinations and must be ruled out with the help of other medical professionals.

### ***Management***

To properly manage a CBS patient, begin with a thorough case history. If patients volunteer their hallucinatory experiences, the clinician should inquire of specific details about the episodes. However, most patients will not openly share such experiences, so it is up to the skilled clinician to identify those at risk for developing CBS and directly ask about hallucinations. Questioning the patient must be performed in a delicate manner. Showing sympathy and being sensitive will likely help make the patient

more comfortable to open up about the subject. For those at high risk of developing CBS but denies hallucinations, one should be the first to inform and educate the patient about CBS. Early patient education will help prepare them for the possibility of developing hallucinations in the future and ease any initial shock or fear when it happens. Continued reassurance and counseling helps ease the worried and fearful patient.

Next, a thorough ophthalmic examination should be performed to rule out ocular and visual pathway pathology. In the presence of some ocular or visual pathway pathology, proper treatment to minimize visual loss and maximize vision is critical. Maximizing visual function with spectacles, visual aids, and/or surgical procedures (i.e. cataract surgery) can help minimize visual deprivation and episodes of hallucinations.

Referring the patient for proper neurological and neuropsychological examinations can help eliminate other potential causes of the hallucinations and prevents misdiagnoses. A referral to psychological therapy for hypnosis, distraction, cognitive restructuring and relaxation training can be of benefit.

Pharmacotherapy has found no universally effective treatment but there are case reports of anticonvulsants and antipsychotics (neuroleptics) helping to decrease the occurrence and frequency of the hallucinatory episodes. Suggesting social and environmental changes which decrease social isolation and sensory deprivation such as establishing support groups and living in nursing homes has been proven helpful to decrease hallucinatory episodes.

## **Conclusion**

CBS tends to occur in patients with some form of acquired vision loss. These patients are often elderly and are more susceptible to ocular diseases that cause visual loss. Patients with CBS tend to either not report their hallucinations or be misdiagnosed when they are reported. As a result, most of them are inappropriately treated and mismanaged. CBS hallucinations can present in various ways which can be easily confused with other conditions. It is important for an eye care provider to be aware of CBS to better identify, diagnosis, manage and refer patients appropriately for the proper care of this condition. Often, the most powerful therapeutic approach for CBS patients is empathy, sensitivity, communication, and reassurance.

## **CONCLUSION of NEURO-OPTO SEMINAR:**

Cortical blindness and Charles Bonnet Syndrome are both commonly found in low vision practices and hospital-based settings. Cortical blindness is characterized as impaired vision as a result of insult to the visual cortex. Visual hallucinations may present with cortical blindness, in what is called Charles Bonnet Syndrome (CBS). CBS is defined as the occurrence of visual hallucinations in intact cerebral functioning patients, secondary to visual deprivation. Therefore, aside from cortical blindness, any form of ocular pathology causing vision loss can cause CBS. It is important for eye care practitioners to be aware of these conditions in order to make confident diagnoses and proper referrals.

### *Cortical Blindness References:*

Alp BN, Bozbuğa N, Tuncer MA, Yakut C. [Transient cortical blindness after coronary angiography](#). J Int Med Res. 2009 Jul-Aug;37(4):1246-51.

Misra M, Rath S, Mohanty AB. [Anton syndrome and cortical blindness due to bilateral occipital infarction](#). Indian J Ophthalmol. 1989 Oct-Dec;37(4):196.

Cortical Visual Impairment." *Cortical Visual Impairment*. Boston Children's Hospital, n.d. Web. 14 May 2013. <<http://www.childrenshospital.org/az/Site2100/mainpageS2100P1.html>>.

Hyvärinen, Lea, MD. "Understanding the Behaviours of Children With CVI." *Understanding the Behaviours of Children With CVI*. American Printing House for the Blind, Inc, Mar. 2004. Web. 14 May 2013. <[http://www.aph.org/cvi/articles/hyvarinen\\_1.html](http://www.aph.org/cvi/articles/hyvarinen_1.html)>.

Bagheri J, Mandegar MH, Sarzaeem MR, Chitsaz S. [Transient bilateral cortical visual loss after coronary artery bypass grafting in a normotensive risk-free patient](#). Heart Surg Forum. 2008;11(4):E248-51

Tanaka M, Jaamaa G, Kaiser M, Hills E, Soim A, Zhu M, Shcherbatykh IY, Samelson R, Bell E, Zdeb M, McNutt LA. [Racial disparity in hypertensive disorders of pregnancy in New York State: a 10-year longitudinal population-based study](#). Am J Public Health. 2007 Jan;97(1):163-70. Epub 2006 Nov 30.

"Faqs." *Faqs*. Preeclampsia Foundation, n.d. Web. 14 May 2013. <<http://www.preeclampsia.org/health-information/faq>>.

Junck L, Marshall WH. Neurotoxicity of radiological contrast agents. *Ann Neurol* 1983;13:469–484

Cunningham FG, Fernandez CO, Hernandez C. Blindness associated with preeclampsia and eclampsia. *Am J Obstet Gynecol* 1995;172:1291–8.

Aldrich, Michael S., Anthony G. Alessi, Roy W. Beck, and Sid Gilman. "Cortical Blindness: Etiology, Diagnosis, and Prognosis." *Annals of Neurology* 21.2 (1987): 149-58. Print.

Saigal G, Bhatia R, Bhatia S, Wakhloo AK. MR findings of cortical blindness following cerebral angiography: is this entity related to posterior reversible leukoencephalopathy? *Am J Neuroradiol* 2004;25:252-6.

Cioni G, Fazzi B, Ipata AE, et al. Correlation between cerebral visual impairment and magnetic resonance imaging in children with neonatal encephalopathy. *Dev Med Child Neurol*. 1996 Feb;38(2):120-32.

Hoyt CS. Brain injury and the eye. *Eye (Lond)*. 2007 Oct;21(10):1285-89.

### Illustrations:

[http://politedissent.com/images/apr06/brain\\_arteries.html](http://politedissent.com/images/apr06/brain_arteries.html)

<http://www.ncbi.nlm.nih.gov/bookshelf/picrender.fcgi?book=neurosci&part=A816&blobname=ch12f2.jpg>

<http://cdn.bmedreport.netdna-cdn.com/wp-content/uploads/2010/10/brain-visual-cortex-study1.jpg>

[http://www.gillesbailly.fr/publis/Large\\_less\\_touch\\_Finger\\_Count.png](http://www.gillesbailly.fr/publis/Large_less_touch_Finger_Count.png)

<http://www.enhancedvision.com/wp-content/uploads/2011/06/cane-300x225.jpg>

### *Charles Bonnet Syndrome References:*

- A. Chaudhuri, Charles Bonnet syndrome an example of cortical dissociation syndrome affecting vision, *J Neurol Neurosurg Psychiatry*, 69 (2000), pp. 704–705
- A. Fernandez, G. Lichtshein, W.V. Vieweg. The Charles Bonnet syndrome: a review. *J Nerv Ment Dis*, 185 (1997), pp. 195–200
- A.M. Levine. Visual hallucinations and cataracts. *Ophthalmic Surg*, 11 (1980), pp. 95–98
- A.M. Santhouse, R.J. Howard, D.H. Ffytche. Visual hallucinatory syndromes and the anatomy of the visual brain. *Brain*, 123 (2000), pp. 2055–2064
- Bonnet C: *Essai analytique sur les facultés de l'ame*. Copenhagen and Geneva, Philibert, 1769, ed 2, pp 176–8
- D.K. Adair, M.S. Keshavan. The Charles Bonnet syndrome and grief reaction. *Am J Psychiatry*, 145 (1988), pp. 895–896
- F. Rosenbaum, Y. Harati, L. Rolak, M. Freedman. Visual hallucinations in sane people: Charles Bonnet syndrome. *J Am Geriatr Soc*, 35 (1987), pp. 66–68
- G.C. Brown, R.P. Murphy. Visual symptoms associated with choroidal neovascularization. Photopsias and the Charles Bonnet syndrome. *Arch Ophthalmol*, 110 (1992), pp. 1251–1256
- G. de Morsier, Le Syndrome de Charles Bonnet hallucinations visuelles des vieillards sans déficience mentale, *Ann Med Psychol*, 125 (1967), pp. 677–702
- Gold K, Rabins PV: Isolated visual hallucinations and the Charles Bonnet syndrome: a review of the literature and presentation of six cases. *Compr Psychiatry* 30:90-8, 1989
- G. Schultz, R. Melzack. The Charles Bonnet syndrome: phantom visual images. *Perception*, 20 (1991), pp. 809–825
- G. Schultz, W. Needham, R. Taylor *et al.* Properties of complex hallucinations associated with deficits in vision. *Perception*, 25 (1996), pp. 715–726
- H. Hécaen, J.G. Badarocco. Les hallucinations visuelles au cours des ophthalmopathies et des lésions des nerfs et du chiasma optiques. *Evol Psychiatr (Paris)*, 21 (1956), pp. 157–179
- H.M. Olbrich, M.P. Engelmeier, D. Pauleikhoff, T. Waubke. Visual hallucinations in ophthalmology. *Graefes Arch Clin Exp Ophthalmol*, 225 (1987), pp. 217–220
- H.W. Kolmel. Complex visual hallucinations in the hemianopic field. *J Neurol Neurosurg Psychiatry*, 48 (1985), pp. 29–38
- J. Damas-Mora, M. Skelton-Robinson, F.A. Jenner. The Charles Bonnet syndrome in perspective. *Psychol Med*, 12 (1982), pp. 251–261
- J.E.A. Bartlett. A case of organised visual hallucinations in an old man with cataract, and their relationship to the phenomena of the phantom limb. *Brain*, 74 (1951), pp. 363–373
- J. Hart. Phantom visions: real enough to touch. *Elder Care*, 9 (1997), pp. 30–32
- Levêque de Pouilly JS: *Eloge de Charles Bonnet*, Lausanne, Henbach, 1794, pp 120–121
- M. Manford, F. Andermann. Complex visual hallucinations. Clinical and neurobiological insights. *Brain*, 121 (1998), pp. 1819–1840

Menon, G. Jayakrishna, Imran Rahman, Sharmila J. Menon, and Gordon N. Dutton. "Complex Visual Hallucinations in the Visually Impaired: The Charles Bonnet Syndrome." *Survey of Ophthalmology* 48.1 (2003): 58-72.

N.J. White. Complex visual hallucinations in partial blindness due to eye disease. *Br J Psychiatry*, 136 (1980), pp. 284–286

Podoll K, Osterheider M, Noth J: [The Charles Bonnet syndrome]. *Fortschr Neurol Psychiatr* 57:43-60, 1989

R.J. Teunisse, J.R. Cruysberg, W.H. Hoefnagels *et al.* Social and psychological characteristics of elderly visually handicapped patients with the Charles Bonnet Syndrome. *Compr Psychiatry*, 40 (1999), pp. 315–319

S. Barodawala, G.P. Mulley, Visual hallucinations, *J R Coll Physicians Lond*, 31 (1997), pp. 42–48

T.L. Schwartz, L. Vahgei. Charles Bonnet syndrome in children. *J AAPOS*, 2 (1998), pp. 310–313

T. Fuchs, H. Lauter. Charles Bonnet Syndrome and musical hallucinations in the elderly. C. Katona, R. Levy (Eds.), *Delusions and hallucinations in old age*, Gaskell, Royal College of Psychiatrists (1992), pp. 187–198