Neurogenesis or the state of continuing creation of ourselves

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ABSTRACT

This syndrome is characterized by the presence of complex hallucinations that are seen by people who have damage along their visual pathway, making them visually impaired. Charles Bonnet Syndrome (henceforth CBS), also known previously as "phantom vision", is the way in which the brain deals with the adjustment of the vision impaired, similarly to how phantom limbs occur within a population who have lost a limb. The brain deals with "loss" via an experience in which it allows the individual to slowly adjust to loss via the release of hallucinations as a compensatory function.

CBS is often seen within the geriatric population and seen across different medical disciplines including optometry, ophthalmology, psychiatry and neurology. Charles Bonnet Syndrome was considered rare before 1990, as there were only a few case histories in medical literature (Sacks, 2012). However, CBS is more common than what was previously reported in literature, and although CBS is still largely unrecognized, even by medical doctors, research suggests that most cases are overlooked or misdiagnosed. A study conducted by Robert Tunisse et al. (1996), who studied six hundred elderly patients in Holland, reported that almost 15% of them had indeed CBS.

Whilst research shows us that the prevalence of CBS is much higher than medical practitioners realize, simultaneously, those who suffer with CBS are afraid of being stigmatized as mentally unstable, and thus many cases go under reported (Kester, 2009). However, creating CBS awareness is of utmost importance more than ever, as we have an aging population and the geriatric population increases. This will also affect the number of those who have acquired vision loss and subsequently cases of those affected by CBS. A study by Gilmour et al. (2009), indicated that only 9% of patients with CBS sought medical advice, and half of them received an explanation on what CBS was. In another study by Cox and Fftyche (2008) observed in an Australian sample of 492 patients in which 47% were not given an explanation on what CBS was, and subsequently had a negative experience from seeking medical advice on the matter. In this study, one third of the medical professionals were uncertain or unaware entirely about CBS. Thus, these indicators are disappointing and as health care providers and researchers, it is important that we recognize and understand this condition in order for these patients to be properly managed and cared for without being misdiagnosed or misinformed.

Keywords: *Charles Bonnet Syndrome, Neuroscience, optometry, ophthalmology, psychiatry, neurology,*

INTRODUCTION (USE "MAIN SECTION TITLE" STYLE FOR MAIN TITLES)

Who is Charles Bonnet

To understand how CBS came to fruition it is important that we understand the history behind the syndrome, starting with Charles Bonnet. Charles Bonnet was a Swiss biologist, philosopher and botanist, and first described visual hallucinations associated with Charles Bonnet Syndrome nowadays. Bonnet was documenting the experience of complex visual hallucinations seen in this 90-year-old grandfather, Charles Lullil and published his findings in his 1760 Essay "Analytique sur les facultés de l'âme" (Sacks, 2012).

His grandfather underwent cataract surgery in both eyes, although his vision was improved, it began to deteriorate over time. The onset of complex visual hallucinations started to occur once he had lost his vision. The initial hallucination began with what was described as a blue handkerchief floating, that followed the movements of his eyes. Anywhere he looked, this handkerchief followed him. The handkerchief over time dissolved and what began to form into a complex hallucination of two men, who wore magnificent cloaks with top hats. This hallucination also dissolved but it was followed by many more hallucinated visitors, all who were reported as being women, who wore beautiful dresses and many who had small boxes on their head. Lullin had an array of hallucinations including incredible flights of pigeons, rotating complex wheels floating in the air, a carriage that grew bigger and bigger and also scenes of men having huge arguments with each other, which to Lullin's amazement, would also follow him in his home, as he saw them fight in miniature form in his living room.

Lullin's hallucinations lasted for several months and then disappeared for good. It's important to note that apart from visual impairment, Lullin did not suffer from any psychiatric disorders (Kester, 2009). Furthermore, Lullin did not suffer from agnosia, as he was perfectly aware that indeed, these visual hallucinations were by no means real. In an ironic turn of events, Charles Bonnet himself, later suffered with vision loss at the age of 20 years old. The vision loss progressed until the age of 40, which by the stage the impairment was severe. He also experienced visual hallucinations like his grandfather. However, the term Charles Bonnet Syndrome, was not coined until 1967 by another swiss scientist, George De Morsier.

What are the Characteristics of Charles Bonnet Syndrome?

Patients with CBS typically have simple or complex hallucinations. The simple hallucinations are often seen in the form of shapes, grid-like patterns or branching patterns, whilst the complex hallucinations are vivid, detailed and contain complicated images of people, animals, faces, vehicles, flowers or trees. Often, what may occur is what is known as a "Lilliputian" hallucination meaning everything will appear in micro-form to what would be expected normatively. Furthermore, it is commonly reported that the figures seen may be pixelated and vary in colors and appearance each time a hallucination is had. In **Figure 1**, an example of what someone with CBS may see as a hallucination is illustrated.

Figure 1. *Representation of a visual hallucination with CBS.*

Source: Wolf (2015). That's Real?!?! – Charles Bonnet Syndrome in Precision Family Eye Care.

http://www.precisionfamilyeyecare.com/charles-bonnet-syndrome/

The hallucination can last a few seconds to hours, with recurrent episodes of visual hallucinations appearing for days or even years. A common trait regarding the hallucinations is that the patients never regard them as disturbing, and mostly will refer to the hallucinations as something funny and bizarre or they will remain neutral about them. Whilst the hallucinations are often referred to as pleasant visual occurrences, it is reported that, at the onset of the hallucination, 39% of patients will be shocked, startled or terrified, but the emotional response will subside decrease up to 8% (Cox & Ffytche, 2014). Often, hallucinations will appear in better vision than what the patient's current visual conditions are. The hallucinations always appear in a clear state of consciousness when patients are alert and awake. It is important to note that in those with CBS, there are no signs of dementia and they maintain intellectual functioning, reasoning, perception and judgment. Often, CBS can be misconceived for early onset dementia. However, it is important to note that CBS is manifested due to a failure of vision and not due to dementia. Additionally. these patients are not agnostic about the hallucination and will always be aware that in fact they are not real, and maintain insight, as by Gilmour et al. (2009), 80% of patients knew that the visual hallucinations were not real after the first episode, followed by 8% by the second episode. Additionally, the images these patients see cannot be triggered or controlled by the patients themselves.

Although often Charles Bonnet Syndrome is seen in geriatric population, there is no age limit for Charles Bonnet Syndrome. CBS has no criteria on age, although only a handful of studies have been conducted on children, which could be a potential area of research for future academics. Thus, to sum up, the features of the CBS hallucinations may be described as the following:

– Color, black or white.

– Static, like a photograph or dynamic, with movement, like watching a film.

– Realistic (people, animals, objects, buildings) or fantastic (mythical creatures).

– Brief (they continue for a few minutes), or lengthy (for a few hours).

– Enjoyable hallucinations, almost like seeing a pleasant dream that are initially frightening.

– Meaningless and random, or featuring familiar people or familiar places.

– Hallucinations are inconsistent and different every time.

– There are no control or triggers for the hallucinations.

An account by a patient with CBS can be seen in the video below known as Bee's Story, a patient with glaucoma who experienced CBS.

Charles Bonnet Syndrome (CBS) | Bee's story

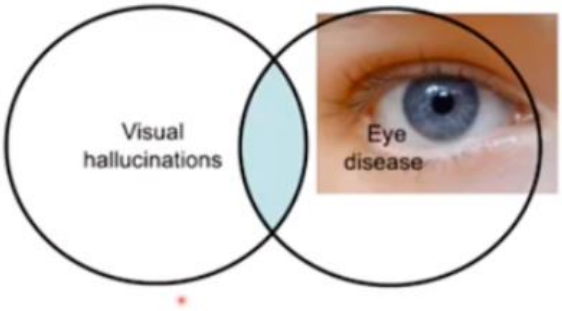
How are visual hallucinations possible in blindness?

As the brain deals with a loss due to impaired vision, as a compensatory technique, the brain continues to maintain visual stimulation. As the eyes itself are not able to function to receive and project visual stimulation, the brain will in a sentence "make the stimulus up". The process itself mimics the same compensatory technique we see in patients with phantom limb syndrome, where patients with a lost limb may still feel it, have sensation of it and feel it tingling even though the limb is no longer there. This is known as a release mechanism which is associated with modality-specific sensory deprivation, otherwise known as Sensory Deprivation Theory. It's important to note that the compensatory technique that the brain attempts in order to make up for lost vision via hallucinations is possible, as the visual system has plasticity and is able to reorganize perceptive fields that can elicit visual hallucinations (Yacoub & Ferruci, 2011). This theory is largely accepted as to why individuals with visual impairment can have Charles Bonnet Syndrome. However, this solely remains a theory as the true underlying mechanism behind the visual hallucinations in CBS are still unknown.

What Populations can Develop Charles Bonnet Syndrome?

CBS affects individuals that have lost more or all of their vision and most likely occurs when impaired vision affects both eyes and the brain is trying to compensate for vision loss. An association has been found with Macular Degeneration, in which it is said that half of patients experiencing it will develop CBS. Figure 2 gives an illustration of the intersection of eye disease and eye diseases, thus hallucinations within eye disease is the confirmed Charles Bonnet Syndrome.

Figure 2. *Intersection between Visual Hallucinations and Eye Disease that confirm CBS.*



How do you Diagnose Charles Bonnet Syndrome?

There is no specific diagnostic assessment for CBS. However, if you have a suspicion that a patient may have CBS you can:

– Talk to the patient about their hallucinations and symptoms in order to understand the hallucinate profile they are suffering from.

– Take a detailed medical history, and rule out any other psychiatric/psychological disorders

– Carry out tests for other possible causations of hallucinations such as Alzheimer's disease.

– Ensure they are not showing any signs of dementia.

– Ophthalmological assessment to be conducted to confirm vision loss that can be indicative that the patient is most likely experiencing Charles Bonnet Syndrome.

Thus, it is important to note that a diagnostic impression of CBS, is based on exclusion. There are many etiologies that can result in having hallucinations. However, an investigation needs to be conducted in order to rule out other syndromes/disorders before concluding to CBS.

What Are the Possible Interventions after a Diagnosis?

In most of the CBS cases, the hallucinations will dissolve on their own between 12-18 months without any specific intervention or treatment. However, letting your patient know and understand what CBS and why it is happening may help relieve and resolve a lot of emotional distress that may accompany an individual who is afraid of being stigmatized as mentally ill. If hallucinations persist after 18 months, and the severity and the intensity of the hallucinations remains, there are some interventions that may be conducted. An optical intervention may be conducted and that is maximizing the vision that the patient has remaining via contact lenses or low vision rehabilitation. Thus, by improving the remaining vision, the frequency of the hallucinations may be reduced (Kester, 2009). If the patient's visual impairment can be treated by surgical intervention, this will completely stop the visual hallucinations, specifically an effective surgical method for treatment of CBS has been laser intervention (Eperjesi & Akbarali, 2004). When these options are exhausted, in severe cases, medical therapy via pharmacology is conducted in order to reduce the hallucinations. Medications such as Venlafaxine may be prescribed in low doses, antipsychotics, anticonvulsants and up taking a serotonin intake may be administered to the patient. Currently, there are no sets of medication that have been created for the purpose of CBS. However, reported cases have used the medical therapy described above. Despite of this, the results of these medications differ by case and patient, and the reduction of the hallucinations has not been generalized (Hartney et al., 2011).

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