The Royal Australian and New Zealand College of Ophthalmologists

Position Statement: Charles Bonnet Syndrome

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Approved by: Board **Version:** Current **Department**: Advocacy	**Next review date:** November, 2026 **Approval date**: November, 2023 **Policy inventory number:**

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We acknowledge the Aboriginal and Torres Strait Islander Peoples, the Traditional Owners of Country throughout Australia and recognise their continuing connection to land, waters and community. We pay our respects to them and their cultures; and to their Elders past, present and emerging.
In recognition that we are a bi-national College, we also acknowledge the Rangatiratanga of Māori as Tangata Whenua and Treaty of Waitangi partners in Aotearoa New Zealand.

1. Purpose and scope

This position statement was developed by The Royal Australian and New Zealand College of Ophthalmologists (RANZCO). The purpose of this position statement is to provide guidance to RANZCO's Fellows and other health professionals regarding diagnosis, treatment and management of Charles Bonnet Syndrome.

Charles Bonnet Syndrome (CBS) is a visual disturbance in which a person with partial or severe blindness, experiences visual hallucinations. People suffering with CBS are aware that their hallucinations are not real. It is not associated with other sensory hallucinations. It is not thought to be related to psychosis or dementia.
To date it has been both under reported[1] and under-recognised both by patients and medical practitioners.[2,3] However, with the formal recognition of CBS as a distinct clinical condition by WHO, (ICD-11 Code 9D56) in 2022, there has been an increase in interest and research into this disease.[4]

The purpose of this review is to increase awareness of CBS amongst eye health care providers, patients, and their carers, so as to decrease the negative impact it has on patients suffering with it.

2. Causes and Characteristics

Charles Bonnet, in 1760, described visual hallucinations suffered by his 89 year old grandfather whose vision had gradually decreased following cataract surgery. These hallucinations were formed images, of people, birds, carriages and buildings. The patient had intact cognition and was fully aware the visions were "fictions of his brain".[1]
The eponym, Charles Bonnet Syndrome, was given to these hallucinations in 1938 by George de Morsier, a French/Swiss neurologist.[5]

However, it was not until thirty years later that the association with visual loss was identified.[5] Typically, CBS is found in older patients suffering from visual impairment, due to pathology anywhere in the visual pathway.[5,6]

CBS can also occur in children with early onset visual deprivation, who have lost sight rapidly. However, it does not tend to occur if the child lost vision prior to the age of four or was born blind.[7,8]

Common causes include age related macular degeneration (ARMD)[9] and glaucoma,[10] but it has been described with visual loss secondary to cataract, corneal opacities, retinal abnormalities (eg retinitis pigmentosa, diabetic retinopathy), and disorders of the optic nerve, optic tract, and visual cortex, including compressive lesions and brain surgery.[11-13] CBS occurs more frequently if the loss of vision is sudden and unexpected.[14] CBS can develop without loss of central visual acuity.[6]

The prevalence of CBS varies depending on the study and whether the hallucination is elementary i.e. phosphene or unformed colours (40-60% incidence) or complex (10-15% incidence).[1] However, the incidence is probably greater, as patients often fail to report the hallucinations due to a fear of being labelled as having a mental illness. Complex hallucinations more frequently affect people with lower visual acuity, more extensive field loss and greater social isolation.[6,14] One study has found hallucinations to be less frequent in Asian populations. The reason for this is unknown.[15]

No one definition of CBS has been universally accepted across the various medical disciplines.[16] It is characterised by vivid, complex and recurrent visual hallucinations in visually impaired people who have normal neuropsychological function and are usually aware that the hallucinations are not real.[13] Some papers suggest that a declining cognitive function may increase the risk of CBS. This, however, is controversial if the definition of CBS is visual hallucinations occurring in a person with normal cognitive function.[1,11] The visual hallucinations occur when the patient is alert and awake and are not associated with other sensory hallucinations.

The classic description of the hallucinations range is of a complex non-threatening formed image of faces, people, animals, buildings, landscapes or other objects.[6] Unformed hallucinations of simple shapes and dots of colour have also been reported.[17] They are perceived to be located in external space, and usually at an appropriate distance and perspective for the patient's environment i.e. indoors or outdoors.[6]

The images usually last for only a few minutes, but sometimes can last for several hours.[18] Recurrences of hallucinations can continue for several months before their frequency starts to decrease. However, in 75% of patients, the hallucinations can persist for 5 or more years.[18]

In cases resulting from intracranial causes, such as hemianopia following brain surgery, the hallucinations tend to resolve over several weeks.[11] Patients who have undergone removal of pituitary tumours often find the hallucinations replaced by photopsias once the tumour is removed.[19]

The neurophysiology of hallucinations is unknown. The more widely accepted theory is hodotropia, or sensory deprivation, where visual loss leads to deafferentation of visual association cortical areas of the brain. This causes disinhibition and later spontaneous activity of the visual regions in the absence of visual input,[14] analogous to phantom limb pain.[20] Another theory is the release phenomenon, where, due to missing visual input to the primary visual cortex areas, a disinhibition of visual association areas occurs, leading to a release of visual hallucinations.[14] Using functional imaging of the cortex, it is hypothesized that the perceived image corresponds to the functions of cortical area active at that time.[1,21]

3. Awareness of CBS

Studies have shown a lack of awareness of CBS amongst medical practitioners and patients. A Canadian study found 55% of GPs were unaware of CBS, and 85% never discussed the possibility of it with their patients.[2] A Danish study of patients attending a retinal clinic found only 12% of patients were familiar with CBS and 36% of patients were not notifying their treating ophthalmologist of their visual hallucinations.[3]

CBS can have a significant impact on patients. Lack of awareness of CBS can increase a patient's anxiety. Cox and Ffytch (2014) found 32% of patients suffering from CBS were negatively affected by the visual hallucinations, due to fear and distress, especially at the time of initial onset. Of the study of 492 patients with CBS, 67% had not heard of the syndrome prior to the onset. 50% attributed the hallucinations to mental illness, with 1/3

afraid of impending insanity, and 2/3 afraid of being labelled as insane if they disclosed the visual hallucinations.[17]

4. Management of CBS

Education of both medical practitioners as well as patients and carers is of great importance.[4,22] Health care professionals, especially those looking after patients with visual impairment, need to be aware of this syndrome. They should openly discuss the possibility of the development of CBS with these patients, especially if the visual loss is due to ARMD or glaucoma. Education of patients and carers should be undertaken, reassuring them that visual hallucinations, should they occur, are not a sign of mental illness or the onset of dementia. However, the development of other neurological symptoms with the onset of hallucinations should prompt referral for consideration of other diagnoses.

If visual hallucinations develop, counselling may be required. This includes the natural history of resolution of the hallucination, albeit after several years. The use of support groups and networking may be of benefit [4], as well as optimising vision as much as possible.

Behavioural exercises may diminish visual hallucinations in some patients. These can include frequent blinking, rapid eye movements, alerting or distraction techniques, or improving lighting levels to improve visual input. Decreasing social isolation and stress levels may also benefit the patients.[23] In patients suffering severe negative impact from visual hallucinations, pharmaceutical treatments with antipsychotics, anticonvulsants, antianxiety medication and SSRI's have been trialled, but with little success.[24]

5. Take home messages for patients and carers

Developing visual hallucinations following visual loss is common, and not a sign of dementia or mental illness.

The frequency and duration of the hallucinations decrease over time, though it may take years.

The hallucinations affect only vision, and do not impact hearing, smell, taste or touch. There is no cure for CBS but being aware of its existence and talking to health professionals, family, friends and carers may help.

CBS hallucinations may lessen or disappear with frequent blinking, moving the eyes rapidly, performing another activity, or changing the lighting level, e.g. switching on a light if hallucinations appear when in the dark.

Try and decrease social isolation and consider joining support groups to meet people who may be experiencing similar phenomenon.

6. Record of amendments to this document

Page	Details of Amendment	Date amended
Entire document Created		2018
Updated and amended entire document		November 2023

7. References

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