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Case Series

# Three cases of Charles Bonnet Syndrome in patients with advanced glaucomatous visual field loss but preserved visual acuity

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**ABSTRACT.**

***Purpose:*** **To describe three cases of Charles Bonnet syndrome (CBS) in glaucoma patients with preserved visual acuity.**

***Methods:*** **Three glaucoma patients who had taken part in a recent CBS study were interviewed about their hallucinations. The patients underwent macular optical coherence tomography (OCT) of both eyes. The visual function was evaluated with visual field measurement (Humphrey visual field analyser) and visual acuity testing (Snellen scale).**

***Results:*** **All three patients had preserved visual acuity (≥0.5 in both eyes) and at least one eye with advanced visual field defect (Mean Deviation worse than −12.00 decibel). They all reported vivid visual hallucinations with insight into the unreal nature of the hallucinations.**

***Conclusion:*** **Charles Bonnet syndrome can occur in glaucoma despite preserved visual acuity. Awareness of this relation is desirable among clinicians, as it will improve communication with patients.**

**Key words:** Charles Bonnet syndrome – glaucoma – preserved visual acuity – visual field defect

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## Introduction

Charles Bonnet syndrome (CBS) is characterized by vivid and complex visual hallucinations in otherwise mentally healthy individuals with visual impairment (Teunisse et al. 1996; Manford & Andermann 1998; Carpenter et al. 2019).

Charles Bonnet first described CBS in 1769 (Bonnet 1769); however, it is still widely mistaken for a psychological disorder due to limited knowledge about CBS among both medical practitioners and patients (Gilmour et al. 2009; Pang 2016). Thus, patients are generally reluctant to report it for fear of being perceived as mentally ill (Pang 2016).

There are various theories explaining the pathogenesis of CBS, but the main hypothesis comprises the deafferentation theory, which suggests that hallucinations are caused by increased activity in the occipital cortex, secondary to reduced sensory input from the eyes (ffytche 2009; Singh & Sorensen 2012).

Charles Bonnet syndrome can affect anyone with decreased visual function (Manford & Andermann 1998; Menon et al. 2003; Tan et al. 2004), but it has also been reported after prolonged blindfolding (Merabet et al. 2004). Many studies suggest that CBS is common among patients with age-related macular degeneration (AMD) (Holroyd et al. 1992; Khan et al. 2008; Vojnikovic et al. 2010; Singh & Sorensen 2012). A recent meta-analysis by Niazi et al. (2020) reported a prevalence of 15.8% in AMD patients.

Charles Bonnet syndrome is traditionally described in patients with low visual acuity (Teunisse et al. 1996; Nesher et al. 2001; Menon et al. 2003; Khan et al. 2008; Gilmour et al. 2009). There are a few reports on CBS in patients with preserved visual acuity (Tan et al. 2004; Madill & Ffytche 2005), and a review and case series by Gold and Rabins (1989) suggests that reduced acuity is not an essential requirement for CBS diagnosis. However, as far as we know, only one previous case report describes CBS in four glaucoma patients with preserved visual acuity (Madill & Ffytche 2005).

In this report, we describe three cases of CBS in patients with advanced glaucomatous visual field defect but

maintained visual acuity. These patients are under clinical care at the glaucoma outpatient department at Skåne University Hospital in Malmö, Sweden. All three patients had taken part in a recent CBS study carried out at the same department and gave their written informed consent to this case report. The patients had undergone automated visual field measurement of both eyes using 24-2 Swedish interactive thresholding algorithm (SITA) programme on the Humphrey Field Analyser (Carl Zeiss Meditec, Dublin, CA, USA). The macula was examined with Swept-Source Optical Coherence Tomography (SS-OCT) (Triton, Topcon, Medical Systems, Tokyo, Japan) at the study visit, and the best-corrected visual acuity (BCVA) was measured using a standard Snellen chart.

All three patients had insight into the unreal nature of their hallucinations and had no neurological (Teeple et al. 2009), psychological (Llorca et al. 2016) or hearing disabilities (Linszen et al. 2019) that could cause hallucinations. None of the patients experienced auditory hallucinations.

# Patient 1

A 73-year-old male patient with bilateral primary open-angle glaucoma since 2014, arterial hypertension and diabetes mellitus. The patient reported no history of serious head trauma (other than a mild concussion 40 years ago).

The BCVA in the right eye was 0.9 and 0.7 in the left eye. The VF of the right eye showed an advanced visual field defect (VFD) stretching from paracentral superior to the nasal and inferior part of the VF (Fig 1). The mean deviation (MD) was −18.00 decibel (dB) and the visual field index (VFI) 44% in this eye. The left eye had an even more advanced VFD with a concentric visual field loss with only a small central island and a minor temporal part remained (Fig 1), a MD of −26.39 dB and a VFI of 19%.

The SS-OCT scanning of the maculae exposed no pathology at all.

The patient had undergone selective laser trabeculoplasty (SLT) in both right eye and left eye a couple of years before and was currently on three-substance ocular hypotensive drug (OHD) for his glaucoma.

This patient reported having intermittent visual hallucinations consisting of colourful paintings, blackbirds flying around his room and children running and playing. He reported seeing hundreds of birds flying around his room in the mornings. He also reported seeing children running towards him. The children would suddenly appear in front of him, making him stop walking in order to avoid stepping on the children, but they would quickly disappear making him realize that it was a hallucination. These hallucinations lasted a few seconds. They came in periods, not every day and were usually not associated with unpleasant feelings.

# Patient 2

An 86-year-old female patient with primary open-angle glaucoma (POAG), senile incipient cataract and arterial hypertension. She had no history of head trauma.

The BCVA in the right eye was 0.8 and 0.5 in the left eye. Both eyes showed advanced concentric VFD with only small central apertures left (Fig 1). The MD was −28.53 dB and VFI 13% in the right eye. The MD in the left eye was −30.27 and VFI 4%. The OCT revealed subfoveal neurosensory detachment in the right eye and drusenoid maculopathy in the left eye.

The patient had undergone SLT treatment in her right eye previously and was on two-substance OHD for her glaucoma.

She reported seeing a woman with long boots walking in front of her. She first experienced this hallucination while taking a walk with her husband. She mentioned telling her husband, how strange it was that the woman had been walking in front of them all the time but found out that her husband did not see any woman in front of them.

The patient also reported seeing the face of a monkey, placed on her grandchild's face, sitting on the sofa in front of her. Other visual hallucinations that the patient reported consisted of an unknown man in a trench coat. The patient also experienced hallucinations consisting of small colourful duvets with grape and clover prints. These hallucinations lasted for many hours during the day, occurring both in- and outdoors, which was disturbing for the patient.

She reported having these hallucinations (the woman in long boots and the face of a monkey) for 5 months until they disappeared in December 2017. However, some of the hallucinations could be transient and intermittent, and they decreased or disappeared during the winter and came back around April when it got lighter. The patient had been experiencing this sporadic reappearance of hallucinations for 3 years in a row. For instance, she had been recently experiencing the hallucination of the man in the black trench coat walking in front of her.

# Patient 3

A 65-year-old female patient with POAG and no history of head trauma.

The BCVA in the right eye was 0.8 and 0.7 in the left eye. The VF in the right eye showed a superior arcuate scotoma and preserved central and inferior visual field (Fig 1). The MD was −11.75 dB and the VFI 72% in this eye. The VFD in the left eye comprised of both superior and inferior arcuate scotoma (Fig 1), a MD of −13.44 dB and a VFI of 66%. No pathology was found on the OCT scan.

She received both Argon laser trabeculoplasty (ALT) and SLT in her right eye and SLT in her left eye many years ago and underwent trabeculectomy in both eyes a couple of years ago. She was currently using a one-substance OHD in her left eye only.

She reported seeing blackbirds, black cats and geometrical structures. These hallucinations started more than 3 years ago. She experienced these symptoms a few times per month. They mostly occurred in the afternoons and lasted a few seconds, sometimes associated with unpleasant feelings. However, she had learned not to pay attention to the hallucinations, which made the experience less disturbing.

# Discussion

The hallucinations in the three cases presented consisted of visual images such as people, animals, plants or paintings – a finding, which is consistent with other studies (Gold & Rabins 1989; Teunisse et al. 1996; Tan et al. 2004).

Charles Bonnet syndrome patients are usually aware of the unreal nature of their hallucinations, but due to the stigma and fear associated with hallucinations and lack of awareness among

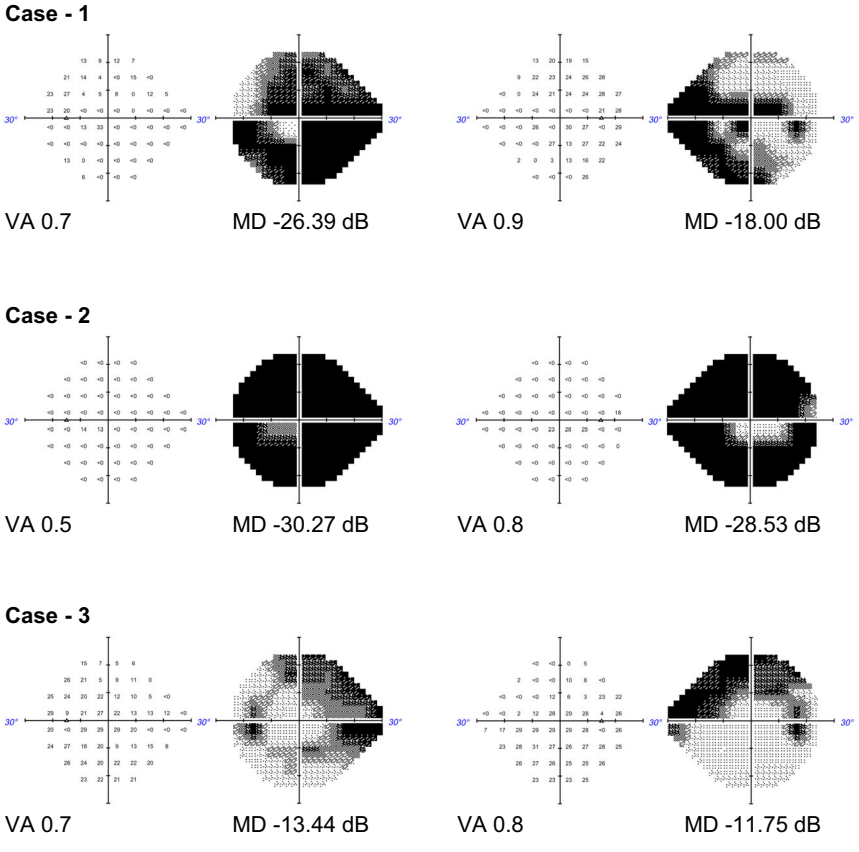


**Fig. 1.** Visual field defects of three patients with open-angle glaucoma and preserved visual acuity reporting CBS symptoms. VA = Visual acuity (measured with Snellen scale); CBS = Charles Bonnet syndrome.

both medical practitioners and patients, CBS is an underreported condition (Pang 2016). Charles Bonnet syndrome (CBS) is also traditionally associated with low vision (Menon et al. 2003; Khan et al. 2008; Gilmour et al. 2009; Singh & Sorensen 2012), which increases the risk of overlooking CBS in patients with maintained visual acuity.

All patients in this report had preserved corrected visual acuity, but advanced to severe visual field defects, which might lead to hallucinations according to the deafferentation theory. These patients were eventually aware of the unreal nature of the hallucinations, but due to the fear of being labelled as dement or mentally ill, the patients in case one and three had not reported having hallucination to their physicians previously. The patient in case two reported severe distress initially as a result of her hallucinatory experiences. She could not contact her own doctor about the hallucinations due to the fear of being regarded to be mentally ill, but she had contacted the department of ophthalmology and talked to a nurse about her experience, without getting any explanation or information on why she might be experiencing these symptoms. She had later searched the Internet for information about her symptoms and mentioned that to an ophthalmologist, who then confirmed her theory about CBS.

## Conclusion

It is important to be aware of the fact that CBS can also occur in patients with advanced glaucoma but preserved visual acuity. CBS is a differential diagnosis of many diseases with the symptom of hallucinations. Hence, it is crucial that not only ophthalmologists, but also other clinicians are aware of this condition. Patients experiencing CBS can initially be frightened as a result of their hallucinations, not knowing why they are having those. They are also afraid to seek help due to the stigma associated with hallucinations. Proper information about CBS and reassurance can alleviate these patients' distress and is thereby an important part of the therapy. Especially in glaucoma patients with advanced visual field defect, ophthalmologists should be aware of CBS in order to be able to give proper information to this group of patients, which can reduce confusion and unnecessary suffering.

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