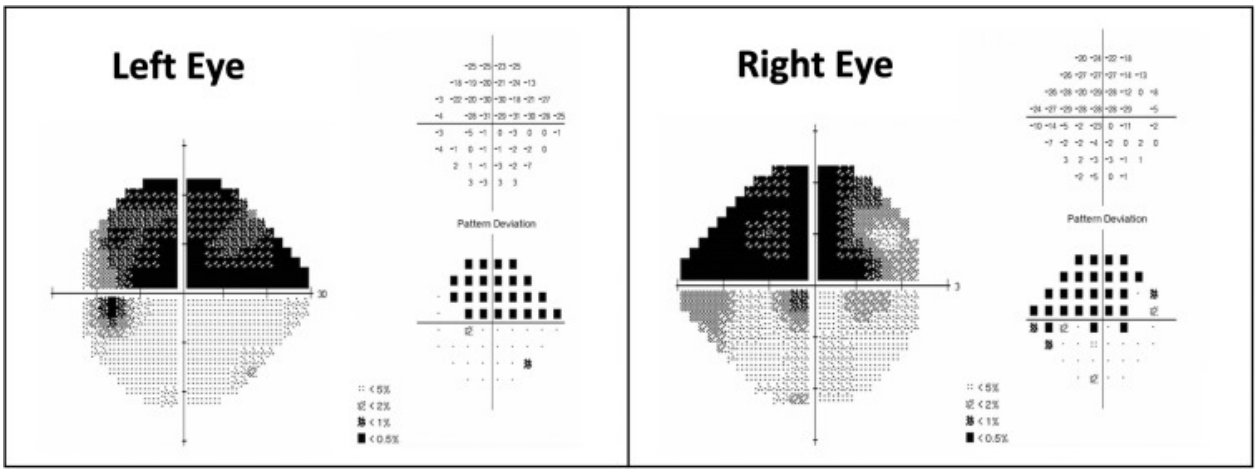
Superior altitudinal hemifield visual hallucinations in Charles Bonnet syndrome



A 79-year-old female was referred to the neuro-ophthalmology clinic for intermittent visual hallucinations. She had a past ocular history of bilateral primary open-angle glaucoma (POAG) and dry age-related macular degeneration (AMD), as well as right eye amblyopia since childhood and strabismus surgery at age 30. Her past medical history included hypertension, essential tremor, bilateral cataract surgeries, and mitral valve replacement. Her medications included ranitidine, dimenhydrinate, bisoprolol, valsartan, rosuvastatin, acetylsalicylic acid, travoprost-timolol eye drops, and vitamins B_6 and B_{12} and folic acid supplements.

The patient had a 12-year history of progressively worsening bilateral superior altitudinal visual field defects from POAG (pretreatment intraocular pressures were 21 and 24 mm Hg, with angles open to grade II–III with moderate pigment), and she reported a more recent 5-year history of episodic visual hallucinations occurring 2–3 times per year. These hallucinations were present even when she had her eyes open, were binocular, and

occurred exclusively in the superior half of her visual field, crossing the vertical midline. The hallucinations included complex imagery, such as a perceived continuation of the wallpaper in a restaurant up onto the ceiling, trees floating in the sky, curtains on the ceiling, and an old house emerging atop a large field. She did not find these hallucinations distressing but rather intriguing, and though they were vivid, she had preserved insight into their nonreality. The patient noticed that fatigue or drowsiness would bring on the hallucinations, but they did not seem to be related to dimly lit environments or social isolation.

The patient's corrected visual acuity was 20/100 OD and 20/20 OS, her intraocular pressures were 12 mm Hg OD and 13 mm Hg OS, and the cup-to-disc ratio was 0.8 OD and 0.9 OS with inferior notches in each eye. Mild macular drusen and atrophy were seen bilaterally, and the patient had a sensory exotropia. Automated Humphrey perimetry revealed dense superior altitudinal visual field defects in each eye with a superimposed subtle generalized depression OD (Fig. 1).

Fig. 1Grayscale and pattern-deviation plots from the patient's Humphrey perimetry showing dense superior altitudinal visual field defects bilaterally.

Computed tomography of the head was unremarkable and showed no lesions in the retrochiasmal visual pathways. Focal visual seizures were excluded on clinical grounds because there was no impairment in level of awareness despite hallucinations involving both the left and right superior visual field quadrants simultaneously. The patient was diagnosed with Charles Bonnet syndrome (CBS), with visual hallucinations confined to her blind binocular superior visual hemifield. The patient declined pharmacologic treatment of her CBS because she was not actually bothered by the visual hallucinations.

CBS was first reported by Charles Bonnet in 1760 when he

describing the case of his own grandfather. The syndrome consists of visual hallucinations occurring in individuals with low vision in which the individual has retained insight into the fact that the hallucinations are not real. The visual hallucinations occur in the absence of other types of hallucinations (e.g., auditory, tactile) and without a neurodegenerative, psychiatric, or pharmacologic cause that would otherwise explain the etiology.

CBS may occur with various etiologies for vision loss, including glaucoma, AMD, diabetic retinopathy, amblyopia, occipital strokes, optic nerve degeneration, and compressive tumours along the optic pathway, and its cause is hypothesized to be the brain attempting to "fill in" areas of sensory deprivation. Because brain regions corresponding to the area of vision loss are no longer constrained by external sensory inputs, their endogenous firing patterns are disinhibited or released, which higher centres perceive as visual hallucinations. For this reason, the hallucinations of CBS are sometimes called *release hallucinations*.

Patients are often reluctant to divulge visual hallucinations to their physicians for fear of being labelled "crazy," and historically this has led to underreporting of the actual prevalence of CBS. However, a recent large study found a prevalence of 39% in a sample of 1254 individuals with macular disease, suggesting that CBS may be much more common than previously thought.[7] Recent reports also suggest that a majority of CBS patients have had visual hallucinations ongoing for >5 years, in line with our patient.

The particularly novel aspect of our patient's CBS was that her hallucinations occurred strictly in the superior hemifield of her vision, where she had dense superior altitudinal visual field defects in each eye from POAG. Previous small patient samples have documented CBS hallucinations occurring exclusively in an area of left or right homonymous hemianopsia due to retrochiasmal cerebral pathology,[7] but on our review of the literature, we believe that our

case is the first report of hemifield hallucinations in CBS respecting the horizontal meridian instead of the vertical.

Our patient had a superimposed pattern of diffusely reduced visual sensitivity in the right eye from amblyopia, and this certainly could have been a risk factor for CBS. However, it did not seem to influence the location of hallucination, likely due to preserved inferior vision in the dominant left eye. The bilateral mild dry AMD also was likely only a minor contributor to her CBS given the noncentral location of the hallucinations and the preserved visual acuity in the nonamblyopic left eye, further supporting that regional and more severe peripheral visual field defects appear to drive hallucination imagery location.

Our patient was taking dimenhydrinate as needed for nausea, a medication that can produce delirium or indirectly contribute to CBS risk or exacerbation through drowsiness. However, our patient remained lucid during her visual hallucinations, and medication effect alone would not account for the strict localization of her hallucinations to her superior binocular visual field.

CBS is an overlooked and underreported phenomenon in patients with visual impairment. Although it is most often recognized to occur in patients with decreased visual acuity, our patient illustrates that CBS also should be considered in patients with dense binocular visual field defects such as can be seen in glaucoma, even in the setting of preserved central vision. Patients are often reluctant to report CBS hallucinations to their doctors, so they should be specifically asked about them. A patient's report of nonmenacing visual hallucinations with preserved insight into their nonreality should alert the clinician to the possibility of a diagnosis of CBS.

Footnotes and Disclosure

The authors have no proprietary or commercial interest in any materials discussed in this correspondence.

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Figures

Fig. 1Grayscale and pattern-deviation plots from the patient's Humphrey perimetry showing dense superior altitudinal visual field defects bilaterally.