

## Case Report

# A Case Report and Review: Charles Bonnet Syndrome Plus With Dementia

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### Abstract

#### Introduction

Charles Bonnet Syndrome (CBS) refers to visual hallucinations in visually impaired patients without psychiatric illness who are typically aware that their hallucinations are not real. Rare cases in the literature describe patients with atypical CBS, or CBS plus, who experience hallucinations in the context of sensory deficits but do not meet all of the criteria of a CBS diagnosis. These cases may include hallucinations in more than one sensory modality, including auditory hallucinations, which are thought to arise by a similar pathophysiology to that of the visual hallucinations in CBS. Unfortunately, the clinical criteria for atypical CBS are ambiguous, potentially explaining the rarity of the diagnosis. In addition, certain features of atypical CBS may make the condition particularly prone to misdiagnosis.

#### Case Presentation

We report a case of atypical CBS in a 67-year-old white male patient presenting with visual and auditory hallucinations that were improved by reassurance. Alongside this case presentation, we provide a review of atypical CBS cases in the literature to compare the diverse features of the syndrome. For this review, we included cases of atypical CBS or CBS plus within the past 20 years for which we could obtain the full text.

#### Conclusion

Clearer guidelines for the diagnosis of atypical CBS and greater attention to the disorder could substantially improve the management of patients presenting with hallucinations. A broader differential diagnosis including atypical CBS for elderly patients with new-onset hallucinations could help clinicians and patients avoid unnecessary medical workup and treatment.

#### Keywords

Charles Bonnet Syndrome; Charles Bonnet Syndrome/diagnosis; Charles Bonnet Syndrome/  
diagnostic imaging; hallucinations/etiology; perceptual disorders; diabetic retinopathy;  
hearing loss; auditory hallucinations; visual hallucinations

#### Introduction

Charles Bonnet Syndrome (CBS) is characterized by complex visual hallucinations in patients with visual impairment who do not suffer from psychiatric illness and are typically aware that they are hallucinating.<sup>1</sup> Atypical CBS, or CBS plus, is arguably less clearly defined, as its delineation has arisen from cases suggestive of CBS that do not fully meet its clinical criteria.<sup>2</sup> Thus, CBS plus can include non-classic features

including cognitive decline, evidence of co-occurring psychiatric illness, and hallucinations of varying sensory modalities.<sup>3</sup> In attempts to characterize hallucinations, further delineations have also emerged. While definitions vary, Musical Ear Syndrome (MES) is thought to be the auditory counterpart of CBS, and the combination of CBS and MES is sometimes referred to as CBS plus.<sup>4</sup>

CBS is associated with a variety of conditions causing vision loss, with studies reporting a prevalence of 10-15% in visually-impaired older adults who have been referred to low vision institutes and ophthalmology clinics.<sup>5</sup> Age-related macular degeneration (AMRD), in particular, is a common cause. A CBS prevalence of 11-26% has been reported in large samples of AMRD patients.<sup>6</sup> However, it should be noted that the true prevalence of CBS is difficult to ascertain due to the possibility of patients underreporting hallucinations. In comparison to that of CBS, the prevalence of CBS plus is even more challenging to establish, especially considering its variable definition (**Table 1**). Though CBS plus is rarely reported in the literature, there are some data on the prevalence of auditory hallucinations that can shed light on the condition. For example, complex auditory hallucinations were reported by 2.5% of patients with severe hearing impairment.<sup>4</sup>

The present report describes a patient with a complex medical history presenting with new-onset auditory hallucinations, requiring consideration of multiple etiologies. We will detail the workup and interventions pursued in this patient with cognitive decline, sensorineural hearing loss, and a mood disorder accompanying his hallucinations. Additionally, a review of recent case reports of CBS plus in the literature, with non-classic CBS features, such as those seen in our patient, will be presented.

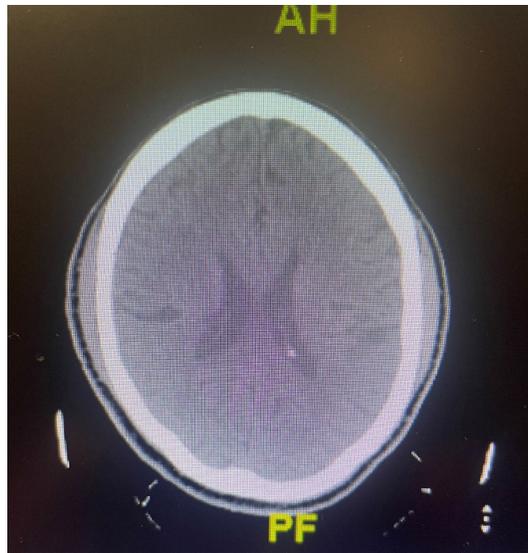
### Case Description

Our patient was a 67-year-old white male military veteran presenting with new-onset hallucinations, which he described as, “I can only hear this unknown person or group of choir members singing songs that are especially bothersome at night. I sometimes see people in the room.” His past medical history was significant for diabetic retinopathy, difficulty hearing due to past trauma (bilateral sensorineural hearing loss), hypertension, hyperlipidemia, major depression, mild neurocognitive impairment, and neurogenic bladder. His wife reported that he was “isolated and [did] not see himself as a useful human being capable of doing things that matter.” On review of symptoms, he reported being “a little depressed” but did not note symptoms of anxiety, post-traumatic stress disorder (PTSD), obsessive-compulsive disorder (OCD), or paranoia and did not display or share other evidence suggestive of thought disorders. He had previously been placed on citalopram for depression and donepezil for memory problems with no improvement in his symptoms for the past few years.

On the mental status exam, the patient appeared alert and oriented, displayed a flat affect, and exhibited poor insight and judgment. He denied any suicidal or homicidal ideation and did not show any signs or symptoms of delusions, paranoia, or auditory hallucinations asking him to carry out certain actions. His past psychiatric history was not significant.

**Table 1.** Comparison of Atypical CBS versus Typical CBS.

	Atypical CBS	Typical CBS
<b>Symptoms</b>	<ul style="list-style-type: none"> <li>Hallucinations in sensory domain(s) other than vision in a patient who has experienced complete or partial loss of the implicated sensory domain(s)<sup>3,4</sup></li> <li>May have visual hallucinations in addition to hallucinations in another sensory domain</li> <li>May have concurrent neuropsychiatric disorder, such as cognitive decline/dementia<sup>1,3</sup></li> </ul>	<ul style="list-style-type: none"> <li>Visual hallucinations in a patient who has experienced complete or partial loss of vision<sup>1,7</sup></li> <li>Full or partial insight into the unreal nature of the visual hallucinations<sup>1</sup></li> </ul>
<b>Exclusions</b>	<ul style="list-style-type: none"> <li>Causes of hallucinations proven to be due to other known pathophysiologic process/disease<sup>1</sup></li> </ul>	<ul style="list-style-type: none"> <li>Primary or secondary delusions<sup>1</sup></li> <li>Hallucinations in sensory domain other than vision<sup>1,7</sup></li> <li>Presence of neuropsychiatric disorder<sup>7</sup></li> </ul>



**Figure 1.** Non-contrast CT of the head, showing diffuse mild atrophy.

Vitals and labs were remarkable for a blood pressure of 131/70 mmHg, glycosylated hemoglobin (HbA1c) of 7.2%, and BMI of 39 kg/m<sup>2</sup>. A non-contrast CT of the head was unremarkable with diffuse mild atrophy (**Figure 1**). Upon neuropsychiatric testing, there was severe impairment on the Rey Auditory Verbal Learning<sup>8</sup> and Trail Making<sup>9</sup> tests, but his performance of activities of daily living (ADLs) was intact. Results were consistent with mild neurocognitive impairment and Patient Health Questionnaire-9 (PHQ-9)<sup>10</sup> was significant for a positive screening of major depressive disorder. The patient was admitted to the inpatient psychiatric facility for his depression, non-command auditory hallucinations, and poor insight. His condition did not improve after the administration of 5 mg of olanzapine. However, with further evaluation and cessation of antipsychotics, reassurance alone began to help him clarify his thought processes and restore his typical mental status. His medications for memory and depression were continued, as cessation would likely have contributed to a further altered mental status in light of his baseline level of cognition, and he was followed up in an outpatient clinic. Although he reported further “musical-like” auditory hallucinations with intact reality testing upon interview, he noted that his overall condition had improved since hospitalization.

## Discussion

Our patient’s clinical picture appears to be

consistent with atypical CBS due to his mild neurocognitive impairment, major depressive disorder, and visual and auditory hallucinations in the setting of diabetic retinopathy and bilateral sensorineural hearing loss. The multimodal hallucinations experienced by this patient were an especially unique component of his presentation. The patient’s hallucinations were unresponsive to antipsychotic treatment, but subjective distress improved with reassurance, which has been found to be an effective treatment for both typical and atypical CBS.<sup>6</sup> His reality testing was intact throughout the encounters, and his hallucinations were “musical”; he denied delusions or paranoia. The patient also reported that even with the use of medications for the previous few years, there had been minimal improvement in his “music-like” hallucinations. In this case, cessation of antipsychotics and reassurance were shown to improve his overall outcome. Other methods that may have helped in this patient’s case are exercise, yoga, and mindfulness techniques to reduce stress and anxiety, which have proven to be helpful in previous atypical CBS scenarios.<sup>6</sup>

## Pathophysiology

While the exact pathophysiology of CBS is not yet clear, several theories are well-recognized. The release phenomenon theory likens the condition to phantom limb syndrome, proposing that the absence of afferent sensory input reduces cortical inhibitory activity, thus causing increased neuronal activity.<sup>11</sup> This increased

neuronal activity is thought to induce hallucinations in the affected sensory system (eg, through amplification of cortical areas normally involved in imagery or music perception). The deafferentation (sensory deprivation) theory suggests that decreased sensory input increases neuronal excitability, allowing small amounts of stimuli to excite neurons and elicit hallucinations.<sup>2</sup>

### Evaluation and Diagnosis

A limited number of atypical CBS cases have been reported in the literature, potentially indicating a diagnostic challenge. For this narrative review, a search of the PubMed database was conducted with the terms ("Atypical Charles Bonnet") OR ("Charles Bonnet Plus"), yielding 10 articles. After evaluation of the results by two independent researchers, 8 studies describing 9 cases were determined to represent atypical CBS. Studies were excluded if they did not mention a diagnosis of a form of CBS.

**Table 2** illustrates the characteristics and treatment of the 9 resulting cases of atypical CBS described in the literature. The scarcity of atypical CBS cases may partly be due to patients' reluctance to report hallucinations due to social stigmas, as well as the unclear diagnostic definition of atypical CBS. For instance, while our patient had multimodal hallucinations and major depressive disorder, other cases involve solely auditory hallucinations without psychiatric conditions. Accordingly, a diagnosis of Auditory CBS has been considered in patients without a significant psychiatric history who experience auditory hallucinations after hearing loss.<sup>12</sup> Hallucinations not associated with psychiatric conditions may even extend to the tactile modality, as described in a case from Sarkar et al.<sup>13</sup> Their patient, who had no psychiatric comorbidities, experienced visual, auditory, and tactile hallucinations that were meaningfully connected by their content, but unrelated to their memories or current stressors. The recognition of a continuum of CBS could highlight the underlying causes of rare hallucinations. In a presentation of two case studies, for example, Dinges et al. described patients with insight into their musical hallucinations, which were later found to be caused by pontine lesions that included the auditory pathway.<sup>14</sup> The authors suggest that the deafferentation and release phenomenon theories could be generalized to apply to hallucinations in patients with brainstem and thalamic lesions.

Atypical CBS can occur in association with psychiatric illness, further complicating its diagnosis. For example, a legally blind 80-year-old woman with a history of fire-related trauma experienced a change in her hallucinations following the Black Saturday bushfires in Australia in 2009. The patient reported great stress after the fires, which triggered frightening hallucinations such as "faces beginning to melt." This finding suggests interplay between CBS and acute stress disorder or PTSD.<sup>15</sup> The influence of stressful life events on the nature of the hallucinations experienced can result in misdiagnosis of CBS as a psychiatric condition and subsequent ineffective treatment with antipsychotics. Moreover, atypical CBS tends to affect elderly patients who are experiencing age-related cognitive decline, which can erroneously lead to diagnoses such as Lewy Body dementia or a primary psychotic disorder. It is important to be aware of the presentation of hallucinations in CBS, which occurs solely without delusions or paranoia, versus a psychosis presentation in early dementia, which may present with command auditory hallucinations, delusions, or paranoia.

### Conclusion

Considering the diverse constellations of symptoms that can be present in patients with CBS and CBS plus, knowledge of these rare diagnoses across various specialties is vital for prompt and effective treatment. With underlying causes potentially encompassing age-related decline, psychiatric considerations, and neurological dysfunction, treatment options can vary significantly. Acknowledgment of diagnoses such as CBS plus can help providers prevent the imposition of unnecessary pharmacological treatment on their patients when simpler interventions such as reassurance may be more effective. Our patient required the continuation of some psychotropic medications for maintenance of his baseline cognitive status, but the introduction of new, intensive pharmacotherapy did not prove to adequately treat him. More moderate, longer-term counseling with such patients may also include recommendations for stress management, such as exercise, yoga, and mindfulness techniques.

The specific criteria for CBS itself are continually under debate, with CBS literature susceptible to overgeneralization when based on few

**Table 2.** Review of Atypical Charles Bonnet Syndrome Cases.

Age, Gender	Authors	Symptoms	Treatment and response
85, F	Abdel-Aziz et al. <sup>16</sup>	Auditory hallucination limited to a repertoire of songs Past medical history (PMH): temporal lobe epilepsy, depression, severe hearing loss, tinnitus	Reassurance (patient retained insight into hallucinations) Patient had learned to control hallucinations by “distracting herself” from them
82, M	Abdel-Aziz et al. <sup>16</sup>	Visual and auditory hallucinations PMH: macular degeneration, deafness	Reassurance
69, M	Jackson et al. <sup>17</sup>	Visual hallucinations PMH: macular degeneration, type 2 diabetes mellitus (DM), hypertension (HTN), arthritis, coronary artery disease (CAD), benign prostatic hyperplasia (BPH), gout	Reassurance
72, F	Arun et al. <sup>2</sup>	Auditory and visual hallucinations PMH: diabetic retinopathy, HTN	Cataract surgery: complete resolution in 1 week
75, F	Dinges et al. <sup>14</sup>	Auditory hallucinations PMH: atrial fibrillation, pontine cerebrovascular accident (CVA)	Pregabalin: rapid improvement
84, F	Van Ranst et al. <sup>4</sup>	Auditory and visual hallucinations PMH: macular degeneration, neurosensorial hearing loss, CVA	Ophthalmologic follow-up Reassurance for auditory hallucinations
86, F	Aziz et al. <sup>11</sup>	Auditory and visual hallucinations PMH: hearing impairment	Olanzapine: partial reduction in auditory hallucinations Carbamazepine: initial visual hallucinations resolved Company and care in nursing home (reduced isolation): both types of hallucinations resolved
86, M	Aziz et al. <sup>11</sup>	Auditory and visual hallucinations PMH: left ear deafness, detached left retina	None reported
70, F	Sarkar et al. <sup>13</sup>	Auditory, visual and tactile hallucinations with insight PMH: bilateral cataract with visual impairment	Clonazepam & reassurance: 50% improvement in symptoms Addition of aripiprazole: complete resolution of hallucinations

case studies.<sup>5</sup> Such generalizations may cause providers to overlook the effect of comorbidities on hallucinations, making it even more important to highlight cases such as our patient’s, which present more atypically. The difficulty in diagnosing atypical CBS in this case demonstrates the importance of further research to establish clear diagnostic criteria.

**Conflicts of Interest**

The authors declare they have no conflicts of interest.

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