

## Abstract

Charles Bonnet (CBS) and Oliver Sacks Syndromes (OSS) are rare neuropsychiatric syndromes associated with deafferentation hallucinations. They manifest as psychotic symptoms of perceptual disturbances in individuals with aging related visual- and hearing-impairment, respectively. Due to the general unawareness of these pathologies among many healthcare workers, patients face barriers when they need to seek help for the evaluation and whether they require interventions. The stigma associated with mental health that is pervasive in our society further adds to the barrier. Herein, we juxtapose the cases of these two fascinating pathologies arising from presbyopia and hypoacusis in the natural course of human aging, in the attempt to raise awareness and educate patients, family, and clinicians and to provide basic guidelines for physicians in identifying, diagnosing, and treating these patients.

## Introduction

Charles Bonnet (CBS) and Oliver Sacks Syndromes (OSS) are rare neuropsychiatric syndromes associated with deafferentation hallucinations. They manifest as psychotic symptoms of perceptual disturbances in individuals with aging related visual- and hearing-impairment, respectively. Due to the general unawareness of these pathologies among many healthcare workers, patients face barriers when they need to seek help for the evaluation and whether they require interventions. The stigma associated with mental health that is pervasive in our society further adds to the barrier. Herein, we juxtapose the cases of these two fascinating pathologies arising from presbyopia and hypoacusis in the natural course of human aging, in the attempt to raise awareness and educate patients, family, and clinicians and to provide basic guidelines for physicians in identifying, diagnosing, and treating these patients.

### Charles Bonnet Syndrome

The visual perceptual disturbance in CBS is thought to be caused by damage to the visual pathway. Differential diagnoses of CBS include but are not limited to intracranial tumors, migraine aura, Parkinson's disease, medication side effects, substance use, and psychiatric causes. CBS is a diagnosis of exclusion and differs between specialties, however consistently recognized is the presence of visual hallucinations in those with impaired vision, intact cognition, and without psychiatric cause. Patients with CBS preserve reality testing and have insight that the hallucinations are not occurring in actuality. Patients may still find these symptoms distressing, however an estimated 1/3 of medical professionals are uncertain of or unaware of this disorder, and many patients do not report symptoms in fear of being seen as mentally unstable. As such, prevalence is estimated at 12-27% of visually impaired patients. Treatment is multidisciplinary and often involves utilization of antipsychotics, anticonvulsants, benzodiazepines, and/or SSRI's.

## Case 1

Ms. T is a 72-year-old black female without previous psychiatric history, who presented to the adult outpatient psychiatry clinic (AOPC) with a 6-year history of isolated visual hallucinations of seeing demons. The patient was legally blind in both eyes from glaucoma and had medical comorbidities such as atherosclerotic cardiovascular disease, hypertension, diabetes mellitus along with the evidence of chronic ischemic small vessel disease on head computed tomography. Although simple hallucinations are most common with CBS, complex hallucinations may occur as in this patient who developed more intricately detailed hallucinations of demons over time. Interestingly in this patient, manifestation of additional preceptory disturbances presented for her auditory and tactile senses. The auditory hallucinations were newer onset and characterized by sounds of drums and bells that became louder as demons came closer and were perceived louder in the patient's deafer ear. The patient also experienced demons crawling on and burning her skin with their fiery spit, which was hypothesized as tactile hallucinations secondary to the patient's peripheral neuropathy.

Although Ms. T's ophthalmologist had already diagnosed the patient having CBS a year prior to her presentation to AOPC, the patient's clinical presentations were inconsistent with CBS, if not atypical, in terms of her development in perceptual disturbances outside the isolated visual hallucinations attributed to her ocular pathology. The patient later also reported delusions of demons entering her bodily orifices, lying in her bowel, and causing her to have constipation. The patient additionally reported hearing the demons hiss when they crawl around her bedroom walls and ceiling like a snake. The patient had poor insight that the hallucinations are not real, and the severity of her symptoms fluctuated with the level of stress or distraction. Patient was treated with quetiapine and clonazepam with CBS, psychotic disorder due to another medical condition with hallucinations, and unspecified schizophrenia spectrum and other psychotic disorder as differential diagnoses, and after a successful titration of the antipsychotics to 100mg nightly, patient's hallucinations and distress have significantly reduced.

## Case 2

Mr. H is an 88-year-old married white male without previous psychiatric history who was admitted to the hospital for a planned transcatheter aortic valve replacement (TAVR). His surgery and post-operative recovery were uncomplicated. During the hospitalization, the patient and spouse reported new onset auditory hallucinations that had begun about 1 to 2 months prior to the TAVR. The patient described intermittently hearing radio music while others could not in multiple settings. Mostly he heard songs, such as those by Johnny Cash or church music, and rarely, he heard radio DJs doing skits or commercials. These auditory hallucinations were most prominent at night and in settings of low background noise, especially when walking to go to the bathroom overnight even though they could come on at any time throughout the day and could last for hours. The patient had insight that his symptoms were hallucinations after realizing that his wife could not hear the sounds and by confirming that the musical volume did not muffle by his covering of ears or removing his hearing aids.

The patient expressed possibly delusional content, namely implying that he thought to be able to call the radio station and request to turn off, if only he knew which station it was. Per wife, the patient has had a gradual, mild decline in his cognition for the past year, specifically impairments in memory and instrumental activities of daily living. He also noted having been involved in a motor vehicle collision about two months prior to this hospitalization, however without acute changes in his functioning. Importantly, Mr. H reported gradual onset progressive hearing loss for which he was prescribed hearing aids last year.

Unlike Ms. T in Case 1, Mr. H was not particularly bothered by the auditory experiences, and in fact, spoke of them fondly at times. Patient reported getting his usual 9 hours of sleep and being able to complete his daily activities despite the intermittent MH, which was isolated without additional perceptual disturbances. His concern was rather that he might be "going crazy" and that neither of them could understand or explain this ongoing phenomenon. The inpatient psychiatry consult team provided psychoeducation targeted at normalization of the symptoms, diagnosis, etiology, and prognosis. The patient and family expressed profound relief from the fear of the unknown by this simple identification and labeling with a diagnosis. In addition, the psychiatry team provided guidance regarding possible therapeutic interventions, such as referral to outpatient ENT and Neuropsychology, and medications. The patient made an informed decision not to pursue medication treatment or following up with AOPC.

### Oliver Sacks Syndrome

Musical hallucination (MH), also known as OSS, is a rare heterogeneous phenomenon that occurs in 0.16% of patients in a general hospital setting and 2.5% of elderly patients with hypacusis. Patients with MH perceive complex sounds consistent with their past musical memories (e.g., carols, musicals, opera) in the absence of an external auditory stimulus. Most cases occur as a seemingly isolated phenomenon without new onset of psychiatric symptoms or cognitive deficits; however, are often multifactorial in etiology. MH is more prevalent in females and the elderly, with the single greatest risk factor being moderate to severe acquired hearing loss; present in 61% of patients with MH. Low background noise, social isolation, and nighttime appear to exacerbate symptoms, and the phenomena is conjectured to be due to "perceptual release" of memories that are normally inhibited by a sustained level of sensory input. This theory is further supported by the fact that nearly all cases of MH are familiar musical sequences. OSS may not involve overt hearing loss and instead result from epilepsy, focal brain lesions, psychiatric disorder, and intoxication/medication as primary etiology. Like CBS, the visual counterpart to MH, patients with OSS often have intact insight and may or may not find the hallucinations distressing. When bothersome, the hallucinations can be loud and intrusive, affecting the patient's daily activities and interfering with sleep. Treatment depends on identifying and addressing the reversible underlying etiology with hearing aids being the most published successful treatment. When no identifiable cause of MH can be identified or properly addressed, medication management with acetylcholinesterase inhibitors, antiepileptics, antidepressants, and may be helpful.

## Discussion

CBS and OSS are sibling deafferentation phenomena associated with isolated visual or auditory hallucinations from presbyopia and hypoacusis, respectively, that are often underreported and underrecognized. They remain largely misunderstood by the general population and healthcare workers alike, leading to unnecessary stigmatization, misutilization and underutilization of medical resources. Additionally, these neuropsychiatric syndromes commonly affect elderly populations already vulnerable to stigma and neglect. Thus, the objective of this abstract is to educate patients, family, and clinicians by presenting cases of CBS and OSS in elderly patients with progressive visual or hearing loss, and despite months to years of hallucinations, who either delayed reporting symptoms due to unwarranted concerns of developing a stigmatized psychotic illness or delayed in receiving medication treatment due to possible misdiagnosis. These cases continue to underscore the challenges surrounding CBS and OSS, even though patients may easily achieve symptom remission with or without medications and simple psychoeducation may provide significant relief for the affected patients and families in distress. The low-cost and low risk screening to elicit relevant history in select populations with specific sensory impairment may yield these potential high benefits, and the dire need to increase clinician familiarity with deafferentation hallucinations calls for consult-liaison psychiatrists to make a significant impact again in both inpatient and outpatient settings.

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## Disclosure

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