

*Case presentation*

## CHARLES BONNET SYNDROME

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The cases of three elderly women with Charles Bonnet Syndrome are reported. Review of the literature includes description of the syndrome, its aetiology, management and association with ocular pathology. Bahrain Med Bull 1995;17:

In 1760 Charles Bonnet described the case of his Grandfather, Charles Lullin, who at the age of 78, cataract-ridden but cognitively intact, started seeing coloured, dynamic and highly organised visual hallucinations<sup>1</sup>. Flournoy<sup>2</sup> published a complete account of the visual hallucinations as dictated by Lullin himself at the age of ninety. Figures of men, women, birds, carriages etc would appear and disappear, would approach or recede; increase or decrease in size, but sounds would never be heard.

Morsier<sup>3</sup> was the first to use the term "Charles Bonnet Syndrome" (CBS) to identify visual hallucinations in the elderly with preserved intellectual functions. As Berrios<sup>4</sup> points out, the eponym has been used to describe "any state of visual hallucinations in the elderly irrespective of accompanying symptomology". He believes this is an over inclusive definition and prefers Bartlett's<sup>5</sup> suggestion that no concurrent psychiatric or cognitive disturbance be present.

Although as a rule, no sounds are made by visions, a case of Charles Bonnet Syndrome has been reported associated with musical hallucinations<sup>6</sup>. The syndrome has also been described in a patient at the age of 387. In addition it has been precipitated by bereavement<sup>8</sup> and, although the condition may resolve when end stage blindness is reached<sup>9</sup>, the condition has also been seen in association with blindness<sup>10</sup>.

In 1987 Norton et al<sup>11</sup> studied 434 consecutive patients referred to a psychogeriatric unit, and described eight cases of visual perceptual disorders resembling the Charles Bonnet Syndrome with three significant findings for the eight patients; the preponderance of females to male patients (7:1), the presence of eye pathology with impaired vision in all eight patients, and a mean age of 83.8 years.

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In another study Olbrich et.al<sup>12</sup>, assessed 43 patients with some visual impairment due to bilateral eye disease for visual hallucinations, and found that five out of the 43 patients merited the diagnoses of Charles Bonnet Syndrome. He found strong indication that sensory deprivation was an essential pathogenetic factor for this phenomenon.

Berrios and Brooke<sup>13</sup>, prospectively examined 150 patients referred for out-patient evaluation and found visual hallucinations in 29%. However, only two patients met their criteria for the CBS.

In general, several cases of CBS have been reported in the literature, yet it is still often overlooked or misdiagnosed and hence under reported. Patients add

to this difficulty as they are often reluctant to recount a history of hallucinations, even on questioning lest they should be considered insane.

#### THE CASES

Case 1: MMH a 73 year old widow, with bilateral cataract. Visual acuity was hand movement for the right eye and 6/24 left eye. She was living with her divorced daughter and a maid.

At interview she was fully conscious, well-oriented for time, place and person, and had a very good memory for both recent and remote events; recollecting the name of the ophthalmologist she saw six months ago as well as recounting events that happened few days ago.

She presented with multiple somatic complaints, and also reported visual hallucinations of women wearing black and seeing men dressed in traditional Arab thobes. The visions appeared and disappeared. She saw the same visions as she visited the ophthalmologist in his clinic. There were no associated voices, and the patient was aware that the "people" were not truly present. Other than visual hallucinations the psychiatric and neurological examinations were normal.

The patient was on Soluble Aspirin (Disprin) 100 mg and Tenormin (Atenolol) 50 mg for hypertension that she had had since she was 65 years old.

The patient was placed in a trial of Melleril 25 mg Bid with resolution of the visual hallucinations. When Melleril was stopped, the visual hallucinations recurred.

Case 2: MAA, a 68 year old lady with bilateral cataract, visual acuity was 6/12 both eyes. The patient had lived alone for nearly fifteen years, but had to move to her married daughter's home following the visual hallucinations which she developed one year previously.

At interview, she was fully conscious, neatly dressed and carried a very animated conversation, responding to questions with ease and appropriateness. She was well oriented for time, place and person. Her only complaint was of "visions" of little people, males and females, who came out from under the carpet, from cracks in the wall, peeped out from the radio and even came down with the running tap water. They did not speak to her, but used sign language to communicate with each other. They would often come close to her especially when she prayed, and would cross in front of the prayer mat. They never harmed her and she felt no particular fear from them, but they made it difficult for her to concentrate on her prayers. The images would disappear when she closed her eyelids. The patient was fully aware that they were not real, but on many occasions she felt they might be "genies" of some sort. However they would still be there when she read verses from the Holy Quran, which made her doubt that they were genies, since they were expected to disappear on reading the Quran. The patient otherwise had no psychiatric or neurological disorder. She was given a trial of Melleril 25 mg Bid, increased to 25 mg tid over three months, but this affected the "visions" only a little. Melleril was later replaced by Carbamazepine 100 mg tid, with a very good result. At follow-up, six months later, the patient reported seeing the "visions" on very rare occasions, but she could live with that.

Case 3: FHY, 75 year old widow with bilateral cataract. She could hardly see a hand movement. She lived with her youngest son, wife and three children.

Patient presented with generalised body ache, fatigue and occasional insomnia which was treated with 2 mg Diazepam PRN prescribed by her family physician. She also complained of seeing visions of small people, a well-formed and clear

picture of a cow, and occasionally a horse nodding its head up and down. Her son challenged her ability to see these things when she could hardly see, but she insisted during the interview that the images were quite clear and she could see them over the blurred vision of her cataracted eyes. The images disappeared on closure of her eyes. The patient stated that she was never frightened by the images, but would occasionally get annoyed with the horse as it had a habit of standing in front of her when she prayed. There were no associated voices and the patient agreed with her family that these "visions" were not real. However, she could not explain why she saw them, and insisted that she saw them as clear as she would have seen them before her eyes became cataracted.

At interview she was fully conscious, well-oriented in time, place and person, and had a good memory. She said that the visions were not constant; they would appear and disappear in a fraction of a second, but that they were experienced mostly during prayer time. The patient had had the visions for five years following the death of her husband, and told her family physician about them. He prescribed Melleril 25 mg at night but to no avail.

Psychiatric examination revealed no depressive illness or grief reaction. Neurological examination was uneventful.

#### DISCUSSION

CBS is the only example of visual hallucinations occurring in the absence of organic or psychiatric pathology and accompanied by insight<sup>14</sup>. The presence of insight however, raises the question whether the reported "visions" should be considered hallucinations since lack of insight is an essential characteristic of a hallucination<sup>1</sup>.

The visual hallucinations in CBS characteristically occur in a state of clear consciousness, are not accompanied by any other disorder of sensory perception, and co-exist with normal visual perceptions with the individual retaining the ability to differentiate his/her normal perception from the hallucinations<sup>1</sup>. The content of the visual hallucinations is well organised and defined, as opposed to the normally blurred perception of real objects in patients with ocular pathology.

Peripheral ocular pathology, central neuro-physiological disturbances, and psychological mechanisms are the main factors that have been implicated in the pathogenesis of CBS with specific significance being placed on ocular pathology<sup>1</sup>. The fact that CBS was found in patients with retina lesions as well as in a number of patients with opacity of the corneas, lens or vitreous, may be regarded as an indication that sensory deprivation is the relevant pathogenic factor associated with ocular pathology<sup>1</sup>. The cause of visual hallucinations in the presence of decreased visual acuity is unknown, but it is likely that sensory deprivation is the cause. Three similar phenomena have been described. The first is the visual hallucinations confined to a specific area of field defect<sup>15</sup>. The second is the visual hallucinations following cataract operations at the beginning of the century, which is attributed to the common practice at that time of post-operative masking of both eyes following cataract extraction. The third is the phantom limb when pain is felt in the amputated part after a limb amputation, which is thought to be a hallucinatory phenomenon of the thalamus<sup>5</sup>.

It seems likely that a similar mechanism operates in CBS. In support of this, Heron et al, have shown that after prolonged visual deprivation followed by return to normal environment, humans experience distortions in object size, shape and colour, as well as apparent movement of stationary objects<sup>16</sup>.

The central neuro-physiological disturbances are regarded as another important pathogenic factor. It is thought that CBS occurs mainly in older subjects due to the predominant occurrence of cerebral disturbances in old age<sup>1</sup>. Vigilance is frequently disrupted in older people and may cause various hallucinatory phenomena<sup>17</sup>.

Another type of hallucination with which ophthalmologists occasionally come across, is that in which visions are experienced in the area of a visual field defect as a result of certain brain disorders<sup>18</sup>. Patients with lesions involving the visual association cortex may experience organized hallucinations that usually consist of stereotyped images of people, animals or objects<sup>19</sup>.

The psychological theories centre on the role of the psyche, conflicts, wishes and past memories influencing the content form of visual hallucinations<sup>20</sup>.

The exact prevalence of CBS is unknown. Berrios and Brook<sup>4</sup> diagnosed it in 1.3% of patients referred to psychiatrists for visual hallucinations, while Norton et al<sup>21</sup> made a diagnosis in 1.8% of patients referred to the psychogeriatric unit for routine evaluation.

The prognosis of CBS varies with the nature of the visual dysfunction. It is possible to help some patients by restoring sensory input e.g. by cataract removal. On the other hand some patients were observed to have their hallucinations cease as they progressed to total blindness<sup>22</sup>. The same investigators have reported the efficacy of anticonvulsants such as Carbamazepine and Phenytoin sodium, but there are no proven trials of such therapy.

#### CONCLUSION

Our reported cases are all elderly with impaired vision, and had clear visual hallucinations while retaining insight into the "visions". They are all neurologically free and have no hallucinations of other sensory modalities. There was no evidence of any organic, mental disorder, or functional psychosis. All cases illustrate the salient features of CBS, the visual hallucinations of people and animals are all well organized and clearly defined. Patients are not frightened by the visions and recognize the unreality of their "visions".

Physicians should be allowed keep in mind the differential diagnosis of this syndrome when assessing elderly patients with visual hallucinations. It is of concern that the CBS may be overlooked or misdiagnosed. The incidence of CBS is surely higher than currently diagnosed due to lack of knowledge of the syndrome, and the patient's reluctance to reveal the history of visual hallucinations for fear of being labeled insane.

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