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The Charles Bonnet Syndrome: A Large Prospective Study in The Netherlands

A Study of the Prevalence of the Charles Bonnet Syndrome and Associated Factors in 500 Patients Attending the University Department of Ophthalmology at Nijmegen

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Background. The aims were to determine the prevalence of the Charles Bonnet syndrome (CBS) in low-vision patients and analyse possible associated ophthalmic and sociodemographic factors.

Method. A semi-structured interview on visual hallucinations was given to 300 adult low-vision patients and 200 elderly general ophthalmic patients. Positive cases were examined with the Geriatric Mental State Schedule and the Mini Mental State Examination. Diagnostic criteria were as follows: complex, persistent, or repetitive visual hallucinations; full or partial retention of insight; no hallucinations in other modalities; and no delusions. Ophthalmic and sociodemographic data were gathered for all patients.

Results. The prevalence of CBS in low-vision patients was 11%. CBS was significantly associated with an age over 64 years and a visual acuity in the best eye of 0.3 or less. No significant associations with ophthalmic diagnoses, patient sex, marital status, or social circumstances were found.

Conclusion. Our findings support association of CBS with sensory deprivation and advanced age.

Since Charles Bonnet described (1760) complex visual hallucinations in his psychologically normal grandfather, many case reports of 'isolated visual hallucinations' have been published (Podoll et al, 1989). However, systematic research into this phenomenon, which De Morsier (1967) called the 'Charles Bonnet syndrome' (CBS), has been limited. This is probably due to the vagueness of diagnostic criteria and the assumed rarity of the syndrome. Cause and pathogenesis are unknown. Empiric data suggest associations of the syndrome with old age, eye disease, female sex, social isolation, bereavement, and cerebral disturbance (Damas-Mora et al, 1982; Gold & Rabins, 1989; Schultz & Melzack, 1991). In a recent study (Teunisse et al, 1994) of a group of patients with suspected isolated complex visual hallucinations, it proved to be feasible to isolate CBS from other (psychiatric) disorders by the following criteria: the presence of formed and complex, persistent, or repetitive visual hallucinations; full or partial retention of insight; absence of delusions; and absence of hallucinations in other modalities.

This paper presents the results of a search for CBS in low-vision and other ophthalmic patients, by the above criteria. Our aims were to determine the prevalence of CBS in low-vision patients and to analyse possible associations between CBS and visual acuity, ophthalmic diagnosis, age, sex, marital status, and living situation.

Patients and method

Two groups of patients attending the Department of Ophthalmology of University Hospital Nijmegen were studied.

The first group ('low-vision group') comprised patients from the low-vision unit, where visually handicapped patients are referred to be supplied with reading aids by an optometrist. Of 304 consecutive patients older than 18 years of age, 300 gave their informed consent to participation in the study.

The second group ('optometry group'), in which most patients were presumed to have a relatively preserved visual acuity, comprised patients over 64 years old from the optometry unit, where patients are referred for optic refraction measurements. Of 203 consecutive patients, 200 gave their informed consent to participation in the study.

Age, sex, marital status, and social circumstances were obtained from the patients; ophthalmic diagnoses and actual visual acuity
status were obtained from the ophthalmic records. All the patients underwent a self-designed, semi-structured interview by one of three trained interviewers, who decided whether a subject had ‘no’, ‘possible’, or ‘probable’ complex visual hallucinations. Inter-rater reliability was measured by computing a kappa statistic for chance-corrected agreement between interviewers 1 and 2 (36 interviews, kappa: 0.96) and interviewers 1 and 3 (32 interviews, kappa: 0.91). If the outcome was ‘possible’ or ‘probable’ hallucinations, the patient was examined by a psychiatrist using a check-list on hallucinations, the Geriatric Mental State Schedule (GMSS; Copeland et al, 1976) and the Mini Mental State Examination (MMSE; Folstein et al, 1975). The psychiatrist decided whether the DSM-III-R criteria for hallucinations and the criteria for CBS described above were met. The criterion ‘hallucinations are persistent or repetitive’ was applicable if (a) the patient had hallucinated at least once in the past four weeks; (b) the period between the first and the last hallucination exceeded four weeks.

In the ‘low-vision group’, we analysed the possible associations between CBS and age, specific ophthalmic diagnoses, sex, social circumstances, and marital status. A comparison was made between the patients over 64 years of age of the ‘low-vision group’, 33 patients (11%) in the ‘low-vision group’ were found to have CBS. In the ‘low-vision group’, 16 additional patients had experienced complex visual hallucinations which did not meet our criteria. Only one of them had experienced hallucinations in the four weeks before screening: a 95-year-old widower who had a complete lack of insight. In the other 15 patients, the time which had elapsed since their last hallucinatory episode varied from two months to five years. In eight patients, the time criterion was the only reason for not including them as cases. In the remaining seven patients, the hallucinations had been due to delirium.

In the 33 patients, CBS had started from six months to 10 years previously. There was a wide variation in the content of the hallucinations: people, animals, flowers and plants, vehicles, buildings, other objects, and sometimes complete scenes. Only two patients perceived just a single form. The other patients reported at least three different forms.

Apart from a dysthymic disorder in three patients, no psychiatric disorders were found. The scores on the MMSE ranged from 22 to 30 (mean: 26.5). We considered these scores to be fair, taking into account the negative influence of visual impairment on the performance of many patients. As before (Teunisse et al, 1994), it proved to be feasible to isolate CBS from other (psychiatric) disorders by the criteria mentioned above.

The prevalence of CBS in five age ranges in the low-vision group is illustrated in Fig. 1. The prevalences of CBS in samples of the ‘low-vision group’ and the ‘optometry group’, arranged according to age, ophthalmic diagnosis, sociodemographic variables, and visual acuity in the best eye, are compared in Table 1. To overcome bias by age, we analysed the influence of sociodemographic variables on the prevalence of CBS in the ‘low-vision subgroup over 64 years of age’. There was some overlap in visual acuity between the ‘low-vision subgroup over 64 years of age’ and the ‘optometry group’. Therefore, the prevalences

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1. Visual acuity was measured in each eye separately by the Snellen chart, on which rows of letters of different standard sizes are printed. The patients are asked to read the letters from a fixed standard distance. Visual acuity is expressed as the ratio of the distance at which the patient is still able to distinguish the letters to the distance at which a person with normal vision is capable of doing so. Example 1: a patient is able to read letters from a distance of 5 m which normally could be read from this distance; visual acuity is expressed as 5/5 or 1 (normal). Example 2: a patient is only able to distinguish letters from a distance of 5 m which normally could be read from a distance of 25 m; visual acuity is expressed as 5/25 or 0.2.

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Fig. 1  Prevalence of Charles Bonnet syndrome in low-vision patients subdivided into five age groups.
of CBS for different ranges of visual acuity in patients from the combined groups were included in the table.

In the ‘optometry group’, 60 patients had a unilateral low-visual acuity (visual acuity in the worst eye: 0–0.30; visual acuity in the best eye: >0.30). Only one of them was found to have CBS.

**Discussion**

In the literature, we found only two prospective case-finding studies on CBS. Olbrich *et al* (1987) detected five cases of CBS in 43 elderly low-vision patients, whereas Berrios & Brook (1984) found two cases in 150 psychogeriatric patients. In other prevalence studies on complex visual hallucinations in visually impaired patients, the concept of CBS was not used, and no general psychiatric examination was conducted.

Our study was the first major case-finding study on CBS in low-vision patients and the only one which used fixed criteria and a control group. The prevalence of CBS in our low-vision population was comparable with the findings of Olbrich *et al* (1987), while the prevalence in the patients with less visual impairment was in the same (low) range as that reported in the psychogeriatric population studied by Berrios & Brook (1984).

CBS was not rare in our low-vision patients and even quite common in the elderly participants. The assumption in the past that the syndrome is a rarity may be due to the fact that patients are often reluctant to share their extraordinary experiences with others. Their fear that they may be thought to be insane is mentioned in the literature (Olbrich *et al*, 1987). Many of our patients were quite relieved to hear that CBS is a known phenomenon.
and that it is not thought to be related to mental disease.

The results of our study show a clear association between bilateral impaired vision and CBS. A visual acuity of less than 0.3 in the best eye is associated with a considerable risk of developing the syndrome. It is rare for patients with unilateral severe visual impairment to develop CBS, as was shown by our finding of only one case in 60 of such patients. One other study (Holroyd et al., 1992) reported an association between complex visual hallucinations (although not defined as CBS) and bilateral impaired vision.

Our finding that CBS is associated with advanced age supports similar assumptions in the literature (De Morsier, 1967).

The prevalence of CBS differed little among the most common ophthalmic diagnoses found in our investigation population. We may conclude that low visual acuity in itself is more strongly associated with the syndrome than specific underlying eye diseases.

In some case series, women clearly outnumbered men (Gold & Rabins, 1989; Teunisse et al., 1994). In this study, we found the prevalence of CBS to be higher in women than in men, but we could not statistically confirm a sex effect.

CBS is thought to be related to social isolation (Teunisse et al., 1994). Holroyd et al. (1992) found a significant association between complex visual hallucinations and the variable ‘living alone’ in their ophthalmic population. In our study (on a larger population and restricted to CBS), we found a trend in this direction, but could not confirm the hypothesis statistically. The same applies to the possible association between CBS and bereavement.

We conclude that CBS is associated with low visual acuity and advanced age, as two independent factors. The association between CBS and visual impairment supports those theories on the pathogenesis of the disorder in which sensory deprivation has a role (Schultz & Melzack, 1991). A reduction of visual input, however, cannot be the sole cause of CBS, because not all visually handicapped have hallucinations, and CBS has been described in patients with normal vision (Podoll et al., 1989). The fact that advanced age is a risk factor raises the question of which age-related phenomena are the additional causative factors. We shall continue this research in pursuit of an answer.

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**References**


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