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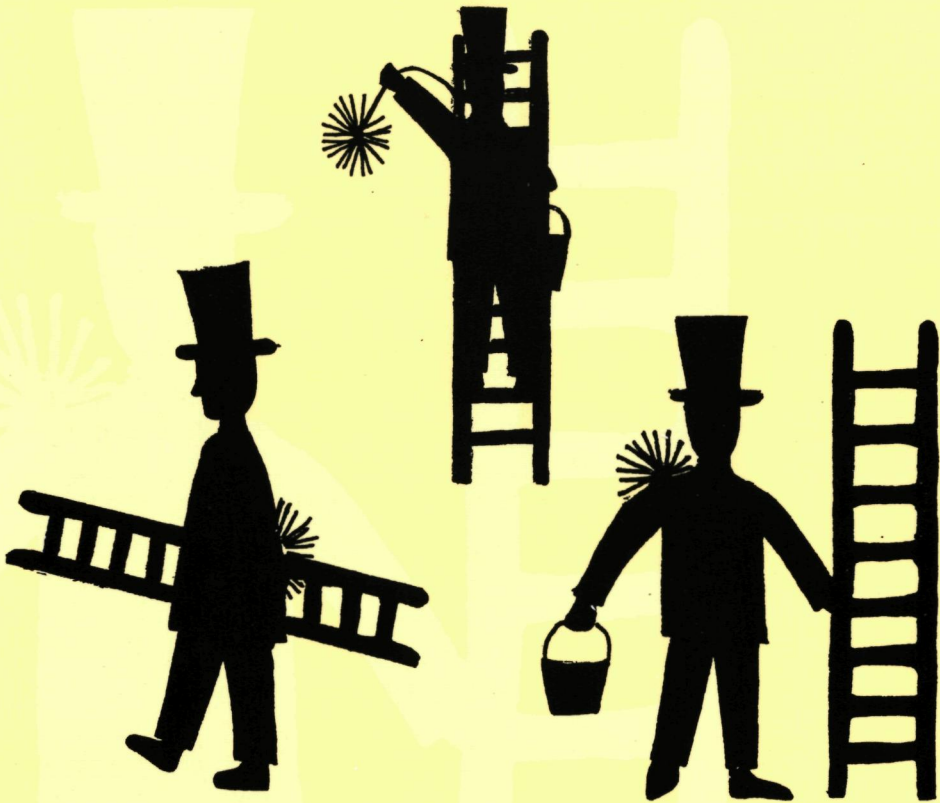
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CONCEALED PERCEPTIONS

Robert J. Teunisse



Concealed Perceptions

An explorative study of the Charles Bonnet syndrome

R. J. Teunisse

Concealed Perceptions

An explorative study of the Charles Bonnet syndrome

Een wetenschappelijke proeve op het gebied van de

MEDISCHE WETENSCHAPPEN

PROEFSCHRIFT

ter verkrijging van de graad van doctor
aan de Katholieke Universiteit Nijmegen,
volgens besluit van het College van Decanen
in het openbaar te verdedigen op
dinsdag 15 september 1998,
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door

Robert Jon Teunisse
geboren op 11 mei 1955
te Ermelo

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Bijziende

*De wereld, waar ik door loop zonder bril,
bestaat alleen uit vlekken en uit vegen.
De vlekken staan voor 't grootst gedeelte stil,
terwijl de vegen allemaal bewegen.*

*Mijn vrienden zeggen: doe toch niet zo dwaas...
zet toch een bril op. Al dat malle turen...
Maar ze beseffen niet, hoe juist dat waas
mij helpen kan het leven te verduren.*

*Ik zie de rozen wel maar niet de luis
en ik zie wel de balk en niet de splinter
en ik zie nooit de rommel in uw huis
en ik zie alles hier en nooit iets ginder*

*Wel zit ik altijd in lijn zeventien
terwijl het twee moet zijn. En wel val ik voorover
de trappen af, omdat ik niet kan zien
waar ze beginnen, maar daartegenover*

*zie ik zo nu en dan een kangoeroe
met een bruin jonkie, op het Leidseplein.
Erg lief is dat, al geef ik later toe
dat het geen kangoeroe geweest kan zijn.*

*Zo staat het dus. En ik ga kippig door
en zeg: hallo Mies, tegen een pastoor...*

Annie M.G. Schmidt

(Uit: De Uilebril. Amsterdam: Em. Querido's Uitgeverij BV, 1988)

This study was conducted at the departments of Psychiatry, Geriatric Medicine and Ophthalmology of the Katholieke Universiteit Nijmegen

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CHAPTER 1

Introduction, aim and outline of the study

R.J. Teunisse

INTRODUCTION

The extraordinary phenomena, we now call hallucinations, have attracted the attention of philosophers, theologians, writers, poets, painters and physicians throughout the ages. In the beginning of western civilization hallucinations were often thought of as supernatural phenomena. This is still the case in some western subcultures and in many non-western cultures. Yet, from early on there were scholars, who tried to understand hallucinations through physiological mechanisms. In more recent times the medical model has prevailed, but hallucinations are still intriguing phenomena and their origin remains a mystery.¹⁻²

The word "hallucinate" is derived from the Latin word "hallucinatus" or "allucinatus" from the Greek origin "hyalein" or "alyein", which literally means "to wander in the mind".² The present meaning of the word "hallucination" is founded on the description given by Esquirol.³ The following modern definition is given in the Diagnostic and Statistical Manual of Mental Disorders, edition 4: "*A hallucination is a sensory perception that has the compelling sense of reality of a true perception without external stimulation of the relevant sensory organ. Hallucinations should be distinguished from illusions, in which an external stimulus is misperceived or misinterpreted. The person may or may not have insight into the fact that he or she is having a hallucination... The term hallucination is not ordinarily applied to the false perceptions that occur during dreaming, while falling asleep (hypnagogic), or when awakening (hypnopompic)...*".⁴

Hallucinations can occur in all sensory modalities, though acoustic and visual hallucinations are the most common types. They can be either elementary (e.g. flashes of light, simple geometrical figures; meaningless noises) or complex (images of people, animals, plants, objects; voices). Complex hallucinations often signal the presence of a severe mental disorder: psychosis. A patient suffering from such a disturbance, may be convinced that the source of the sensory experience is an independent physical reality. He or she may respond emotionally with great anxiety and despair and may act in an irresponsible way upon the hallucinations. Apart from hallucinations psychotic patients often have disorders of thought, such as delusions and confusion.

However, hallucinations are not always symptoms of mental disorder. In 1760 the Genovese philosopher Charles Bonnet wrote the following: "*Je me bornerai donc à dire, que je connois un Homme respectable, plein de fanté, de candeur, de jugement & de mémoire, que, en pleine Veille, & indépendamment de toute impression du dehors, apperçoit de tems en tems, devant lui, des Figures d'Hommes, de Femmes, d'Oiseaux, de Voitures, de Bâtimens, & c. Il voit ces Figures se donner différens mouvemens; s'approcher, s'éloigner, fuir; diminuer & augmenter de grandeur; paraître, reparaitre: il voit les Bâtimens s'élever sous ses yeux..., les tapisseries de ses appartemens lui paraissent se changer tout à coup, ces tapisseries se couvrir de tableaux qui représentent différens paysages..., les hommes et les femmes ne parlent point et aucun bruit n'effleure son oreille... Mais ce qui est très important à*

remarquer, c'est que ce vieillard ne prend point comme les visionnaires ses visions pour des réalités.” (“I just want to say, that I know a respectable man, who is in good health, is reliable, with good judgment and memory, who, in clear consciousness, and independent of all impressions of the surroundings, perceived from time to time, figures of men, women, birds, carriages, buildings, etc. He saw the figures make various movements: approaching, moving away, fleeing, growing smaller and larger, disappearing and reappearing. He saw buildings rise up before his eyes...To him the tapestries of his rooms seemed to change continuously, these tapestries were covered with paintings representing various landscapes...The men and women did not speak and never a noise reached his ears... But it is very important to note that this old man never regarded, as visionnists do, his visions as realities.”)⁵

The old man described by Bonnet was his grandfather, Charles Lulin, aged 89 years. Since 1760 several reports on complex visual hallucinations in mentally normal people, often elderly persons with eye diseases, were published. Honouring the man who was the first to make notice of this phenomenon, the eponym “Charles Bonnet syndrome” (CBS) was introduced by De Morsier in 1936.⁶ Authors discussed the diagnostic criteria, the clinical characteristics and proposed intriguing theories on aetiology and pathogenesis of the CBS.⁷ However, these theories were based only on small case series or even single case studies. CBS was thought to be rare for a long time. In a review of the international literature since 1760, published in 1989, only 46 cases were detected.⁸ Though some authors expressed their doubts on the assumed rarity of the syndrome, little systematic research was done.⁹⁻¹⁰

AIM OF THE STUDY

It is the purpose of this study to analyse the clinical characteristics of CBS and to study connections between CBS and the assumed risk factors that have been mentioned in literature.

OUTLINE OF THE STUDY

In chapter 2 the study starts with a review of the literature on the CBS. Among the different definitions of CBS the most reliable definition is chosen and further operationalized. The literature is screened for proposed risk factors and theories on aetiology and pathogenesis.

Chapter 3 contains a study of a group of patients who assumingly had CBS. The chosen definition of CBS was tried out and the patients were screened for the risk factors proposed in literature with the aim of identifying common factors.

In chapter 4 the prevalence of CBS in the assumed risk group of visually handicapped is determined and associations of CBS with ophthalmic and demographic factors are studied.

In chapter 5 the clinical characteristics of CBS are studied in visually handicapped

patients who were found to have CBS in the prevalence study, aiming to find common features.

In chapter 6 and 7 elderly visually handicapped patients with and without CBS are compared with regard to respectively somatic and psychiatric, and psychological characteristics, aiming to identify risk indicators other than ophthalmic and demographic factors.

Chapter 8 contains a general discussion on the findings of this study and proposals regarding further research.

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CHAPTER 2

The Charles Bonnet syndrome

A review of the literature on isolated complex visual hallucinations

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ABSTRACT

This article presents a literature review on the Charles Bonnet syndrome. By way of illustration, the symptoms are described as encountered in an elderly neuropsychiatrist. The Charles Bonnet syndrome is characterised by the occurrence of complex visual hallucinations in psychologically healthy persons. Knowledge about this syndrome is based almost entirely on casuistry. The syndrome has chiefly been described in elderly people with abnormalities in the peripheral or central visual system. It is likely that the syndrome is not as rare as was previously thought, because many cases will remain undetected as patients are too ashamed to describe their experiences and doctors are unable to recognise the symptoms. Etiology and pathogenesis are unknown. Scientific research into the Charles Bonnet syndrome might provide greater insight into the pathogenesis of hallucinations.

CASE REPORT

Colleague S continued to practice neurology and psychiatry until he was 70 years of age. He was now in his nineties and as a widower led a withdrawn life in a senior citizen's flat. A few years earlier he had decided - in view of his advancing years - only to do enjoyable things. He occupied himself mainly with philosophy and was in the habit of interrogating anyone he met in a Socratic manner. *Colleague S* had never been seriously ill and was not taking any medication. However, his visual and aural capacities had deteriorated, he sometimes suffered from dizziness and he considered himself forgetful.

Five years ago, while having lunch, to his amazement he saw a colourful procession of people with flowery umbrellas entering his dining room. He was looking at them from above, as if he was standing on a platform. The group waved cheerily at him and involuntarily he waved back. Since then, *Colleague S* has had a wide variety of hallucinations, sometimes daily and then sometimes not for many weeks. He saw images of dogs, cows and even more exotic animals, such as wolves and lions, hanging in front of his bookcase, as if there had been a television set there. Various buildings and sometimes complete streets just hung in the air. From his carpet, green stems sprouted that developed into real plants with pink buds. While urinating into the toilet bowl, the water often turned into a colourful street map. He saw all these images just as clearly as if they had really been present, and sometimes even more clearly.

The landscapes, animals and persons he saw were generally unfamiliar to him. Once he saw a person who looked like a caricature of a lady acquaintance. He may have seen some of the animals in nature films, but he was unable to remember precisely.

Colleague S was usually easily able to distinguish the hallucinations from reality: the images did not 'fit' into the surroundings and there was never any noise. However, there were instances that he was less certain. For instance, one day he encountered a man with an oriental appearance lying on his bed. *Colleague S* approached the bed in order to investigate. Even at close range, the man appeared to be real: he was just lying there grinning back. *Colleague S* spoke to him, but the man did not respond. To be on the safe side, *Colleague S* pressed the alarm button, which like many other elderly people, he always carried with him. The caretaker, who arrived promptly, explained that the bed was empty, which was a relief to *Colleague S*. He could still see the oriental gentleman.

Colleague S did not discover any circumstances that encouraged the hallucinations. He was unable to call up the images: they appeared spontaneously and stayed for an average of ten minutes. If he closed his eyes or looked in another direction, he could not see them; but if he turned in the correct direction, then the images were often still there.

It gave *Colleague S* a certain form of pleasure to observe the images. 'I wish I had experienced this while I was teaching the young men and women from nursing about psychiatry', he sighed. He was willing for us to publish his experiences in the *Tijdschrift voor Psychiatrie*.

INTRODUCTION

In the 'Essai analytique sur les facultés de l'âme', published in 1760, the Genevan philosopher Charles Bonnet described the lively and multiform visual hallucinations of his grandfather. The at that time 89-year-old Charles Lullin was in the habit of lecturing his listeners in an amusing manner about the people, animals, plants, buildings and carriages that passed him by on the wallpaper and rugs in his home. Bonnet mentioned explicitly that his grandfather was physically and mentally healthy and was fully aware that the above-mentioned objects were not actually present.¹

Since then, this case report has been presented many times in the scientific literature as an example of hallucinations outside the framework of mental disease. Similar phenomena were reported and De Morsier started to associate the name Charles Bonnet with what was generally considered to be an intriguing rarity.² Over the period 1760 to 1989, Podoll and co-workers encountered a total of 46 case reports in the international literature.³

However, over the past few years, doubt has been cast on the assumed scarcity of the Charles Bonnet syndrome (referred to below as the CBS) by several authors.³⁻⁵ They believe it is possible that the syndrome often goes unrecognised. This formed the reason why we performed a literature study on the CBS. The above-described case report has been added to increase the recognizability of the syndrome.

METHODS

With the aid of CD-ROM and the Medline file January 1981 to December 1994, literature data were collected from articles that contained the keywords *hallucinations*, *pseudohallucinations* and *the Charles Bonnet syndrome*. Articles were selected whose subject, according to the title and/or abstract, comprised the CBS. By means of the reference lists from these articles, other works (e.g. older articles and books) could be traced that were not mentioned by Medline. The following points were abstracted from the resulting literature: psychopathological characteristics, diagnostic criteria, prevalence, risk factors, theories about etiology and pathogenesis and modes of treatment.

RESULTS

Psychopathological characteristics

Rich, multiform and detailed contents of the hallucinations are typical of the CBS: people, animals, plants, inanimate objects and even complete landscapes have been observed. Not seldom, large groups of people are described, such as troops of marching soldiers; miniature people, children and fairy-tale figures such as trolls and gnomes, are also mentioned remarkably often. However, the contents can also

comprise day-to-day issues. The images mostly vary, but sometimes certain objects recur. The objects can be stationary, or show intrinsic movement. It is also possible for an image to shift 'en bloc'. The images sometimes seem to float in the air, or be projected on to a wall, but they can also be integrated realistically into the surroundings. The hallucinating persons put in a silent appearance, while the hallucinated objects have none other than visual qualities.^{3,6-7}

In the literature, there are discussions about whether the hallucinations are new creations or reproductions of images seen in the past. A few patients thought that they could place the objects, whereas many others could not remember ever having seen them in reality.^{3,6-7}

The contents of the hallucinations are not usually frightening. The majority of patients feel emotionally neutral or even amused. Only a small minority feel anxious.⁶ Many authors indicated that the patients realised the unreal character of the images directly or after a short while.^{6,8} However, in some cases the images were so lifelike that the patients had difficulty distinguishing them from reality. They therefore called upon others for reality testing or attempted to discover the truth by means of touch.³

The frequency and duration of the hallucinations vary widely, often even within individuals. Periods of frequent episodes are alternated with periods of absence. The hallucinations can last for a few seconds or up to several hours. Various authors observed that hallucinations occurred particularly when the patient was resting and not concentrating on intellectual work.^{3,7} None of the patients were able to summon the images at will or influence the contents. Some of the patients could make the images disappear by fixing them with their eyes, or by walking towards them. The effect of closing the eyes varied between patients.^{3,9} Norton-Wilson and Munir described a patient who could make the hallucinated person disappear by explicitly asking him/her to do so.¹⁰

The long-term course of the CBS varies: it can be episodic, relapsing or chronic.³

Diagnostic criteria

In the vast majority of articles on the CBS, the following characteristics are mentioned: complex visual hallucinations; photopsia or simple geometric figures are not considered to form part of the syndrome; the patients sooner or later realise that the images are not real; no psychiatric disorders have been established than can explain the presence of the hallucinations. On the basis of these data, Gold and Rabins presented the following diagnostic criteria: (1) the presence of solid, complex, persistent or repeated stereotypical visual hallucinations, (2) full or partial awareness, (3) absence of primary or secondary delusions, (4) absence of hallucinations in other sensory modalities.⁹ In our opinion these criteria are acceptable, with the exception of the condition that they must be stereotypical, in view of the multiformity described in many case reports.

Other diagnostic criteria mentioned in the literature appear to be less valid. De Morsier and Podoll considered advanced age as an inclusion criterion and the presence of a cerebral disorder as an exclusion criterion.^{3,8} However, they did not explain why

an identical phenomenon should be placed in a different diagnostic category exclusively on the basis of age. In relation with the above-mentioned exclusion criterion, Gold and Rabins put forward that not enough progress had been made in scientific research into the CBS to exclude, without reservations, a relationship with a cerebral disorder.⁹ The visual hallucinations described in patients with lesions in the central visual system, such as occipital infarctions, are very similar to those in the CBS and are explicitly included in the syndrome by some authors.^{7,11-12}

Prevalence

No epidemiological research concerning the general population has been performed into the CBS. The results of prevalence studies on certain patient groups are summarized in Table 1.

Table 1. Prevalence of the Charles Bonnet syndrome (CBS) in various patient populations

Study	Population	CBS n (%)
Berrios & Brook (1984) ¹³	150 geropsychiatric patients	2 (1.3)
Norton-Wilson & Munir (1987) ^{10*}	434 geropsychiatric patients	8 (1.8)
Olbrich et al. (1987) ⁵	43 elderly severely visually handicapped patients	5 (11.6)
Brown et al. (1992) ¹⁵	100 visually handicapped patients with lesions in macula and choroidea	12 (12)

* retrospective research

In various other studies the prevalence of complex visual hallucinations in patients with disturbances in the peripheral and/or central visual system varied from 6% to 21%. In these studies, psychiatric examination was limited or lacking. Observations by the authors, such as 'patients seemed to be psychologically normal' and 'patients had no psychiatric history', are strongly suspicious of the CBS.¹⁵⁻¹⁸

Risk factors

It is probable that the syndrome is associated with advanced age and visual disorders. In the above-mentioned overview by Podoll and co-workers of 46 patients mentioned in the international literature, only four patients were younger than 65 years and only four had normal sight.³ The indications were weaker for other factors associated with the CBS. The literature mentions: disturbed acceptance of visual loss, being female, bereavement and social isolation.^{10,15,19-20}

Aetiology and pathogenesis

The cause of the CBS is unknown. A few of the many hypotheses on the origination of the CBS are sketched below.

On the basis of visual disease as the cause of the syndrome, Horowitz hypothesized that the objects were reflections of structures in the eye, such as opacities in the lens or vitreous humour, or reflections of the retinal vascular system. In effect, this would suggest the illusionary falsification of objects that are not positioned peripherally from the eye, but peripherally from the retina.²¹

Guiraud postulated that through pathological stimulation of receptor cells or synapses in the retina, 'meaningless stimuli' are sent to the cerebrum and the cerebrum transforms them into meaningful images.²²

In various theories, loss of sight is considered to be the cause of the CBS. The 'perceptual release theory' suggests that the images incited by sensory stimulation leave behind 'memory traces', but that normal visual input suppresses a person's consciousness of them. As the input decreases, disinhibition occurs and the 'engrams' are once again perceived as images.²³⁻²⁴

Schultz & Melzack applied a theory to the CBS that had originally been suggested by Melzack to explain phantom phenomena. Sensory experiences were thought to be based on nerve-impulse patterns flowing through the wide-spread neural network in cerebro (neuro-matrix). Besides normal sensory stimulation, these patterns can be aroused by non-specific input from the formatio reticularis ascendens, by visual receptors that are still intact, or by hyper-reactivity of neurons in the vicinity of damaged visual centres.^{7, 25}

On a limited scale, electro-encephalography, computer tomography and MRI have been applied to CBS patients. No specific abnormalities were found.^{3, 12, 26-27} Various authors nevertheless believe that the CBS is probably caused by a cerebral disturbance. Such a cerebral disturbance is considered to be too subtle to be detected by existing diagnostic instruments.^{8, 28} Rosenbaum et al. suggested a relationship with temporal epilepsy: a small irritative focus in cerebro might lie at the root of the CBS.²⁹

Flynn presented a psychoanalytical theory: sensory deprivation owing to loss of sight leads to withdrawal of the affective occupation of external objects and the creation of substitute reality through the regressive fulfilment of desire.³⁰

It should be mentioned that no research has been performed to test any of the above-mentioned hypotheses.

Treatment modalities

In the literature, there are various reports on patients who were treated without avail with neuroleptics.^{4-5, 19, 26-27, 31} In contrast, there was only one case with a definite positive response.¹² Carbamazepine was reported to have cured the hallucinations in two patients.^{19, 26} All in all it can be stated that there is little evidence that psychopharmaceuticals are effective.

In some cases the hallucinations disappeared after the patient's sight had been improved, e.g. by cataract extraction.^{5, 32-33}

Although there seems to be little in the way of effective treatment, many authors have indicated the importance of doctors recognising the syndrome. The contents of the hallucinations are not usually frightening, but the fact that he/she hallucinates can be a burden for the patient. Many appeared to have remained silent for a long time about their exceptional experiences out of fear that they would be ridiculed or considered demented. Just by talking to the patient, and possibly his/her relevant others, about the fact that the phenomenon is fairly well-known, even has a name and does not mean that he/she is 'mad', can have an important reassuring influence.^{3,4}

DISCUSSION

Publications on systematic scientific research into the CBS appear to be thinly sown. The lack of generally accepted diagnostic criteria and the assumed rarity of the syndrome are probably to blame for this. The few available prevalence studies give rise to the suspicion that the CBS is more common than was previously assumed, at least in patients with visual disorders. It is therefore important that clinicians are able to recognise the syndrome, even if it is only to reassure the patient and to spare him/her from futile treatment with neuroleptic drugs.

The hypotheses on the etiology and pathogenesis miss an empirical basis. Thorough, controlled, scientific research into the somatic and psychological factors associated with the CBS is necessary to test the hypotheses or to formulate better ones. The importance of such research goes beyond the meaning of the syndrome. The CBS offers the opportunity to study the phenomenon of hallucinations - a symptom for which there is no animal model - in an isolated form. Knowledge about how the CBS originates can also contribute to our insight into the pathogenesis of hallucinations in general.

CONCLUSIONS

Very little scientific research has been conducted into the Charles Bonnet syndrome. Etiology and pathogenesis are unknown. The syndrome is chiefly described in elderly persons with visual disturbances and is probably less rare than was previously assumed. It is important to recognise the syndrome in clinical practice in order to be able to reassure the patient (and his/her relevant others) that he/she does not have a psychiatric disorder. The syndrome offers an excellent opportunity to conduct scientific research into the phenomenon 'hallucinations'.

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CHAPTER 3

Clinical evaluation of 14 patients with the Charles Bonnet syndrome (isolated visual hallucinations)

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ABSTRACT

A group of 14 patients with isolated visual hallucinations who met the criteria proposed by Gold and Rabins for the Charles Bonnet syndrome (with the exception of the criterion 'hallucinations are stereotyped') underwent psychiatric, neurologic and ophthalmologic tests. Additional common characteristics of the hallucinations included: the absence of personal meaning of the content of the hallucinations and the disappearance of the hallucinations when the patients closed their eyes. There was no evidence for a relationship of the syndrome to psychiatric disorders. In the majority of cases, ocular pathology and neurologic disturbances were diagnosed. There was also evidence that most patients were suffering from social isolation. From these data it is concluded that a combination of factors is probably responsible for the Charles Bonnet syndrome.

INTRODUCTION

In 1760, Charles Bonnet described the occurrence of vivid visual hallucinations in his psychologically normal, visually impaired grandfather.¹ Since then, this phenomenon - later to become known as the Charles Bonnet syndrome - has been observed in more patients, with or without clinical evidence of ocular pathology.²⁻¹² Phenomena hardly distinguishable from the Charles Bonnet syndrome have been reported in patients with cerebral disorders and as side-effects of medication.¹³⁻²² However, the accumulation of data about this syndrome has been impeded because most papers are case reports of one or two patients only. In addition, there is a lack of generally accepted diagnostic criteria.

Recently criteria for this syndrome have been proposed by Gold and Rabins:²²

(1) the presence of formed, complex, persistent or repetitive, stereotype visual hallucinations; (2) full or partial retention of insight; (3) absence of primary or secondary delusions; (4) absence of hallucinations in other modalities.

In this report, we describe the psychiatric and medical evaluation of 14 patients who met the criteria proposed by Gold and Rabins, with one exception: the obligatory stereotype nature of the hallucinations. We made this exception because the multiformity of the hallucinations is a particularly impressive feature in many published case histories.^{1,7,10,12,23-25} The aims of our study were to examine the characteristics of the hallucinations described in the literature in our patient population, to establish possible common characteristics and to investigate a relationship between the Charles Bonnet syndrome and psychiatric, ocular, cerebral and metabolic disorders or the use of potentially hallucinogenic medication.

PATIENTS AND METHODS

We asked the physicians of the departments of psychiatry and geriatric medicine of the University Hospital Nijmegen to refer patients with suspected isolated visual hallucinations for our study. A self-developed semistructured questionnaire and the Geriatric Mental State Schedule (GMSS) were used to examine whether the DSM-III-R definition for the concept of hallucination ('a sensory perception without external stimulation of the relevant sensory organ, having the immediate sense of reality of a true perception') and the criteria for the Charles Bonnet syndrome mentioned above were met.²⁶⁻²⁸ (A copy of the questionnaire can be obtained from the authors of this report.)

Patients with the Charles Bonnet syndrome were administered a Mini-Mental State Examination (MMSE) and an elaborate interview by a psychiatrist who classified psychiatric disorders not listed among the exclusion criteria according to the DSM-III-

R criteria, using all data available (GMSS, MMSE, general practitioner [GP] file, hospital file).^{26,29}

Characteristics of the hallucinations were delineated using a checklist. Data pertaining to medical history and medication use were obtained from anamnesis, heteroanamnesis and by reviewing the patient's GP file and hospital file. Vision was tested using Snellen's chart. All patients were examined by an ophthalmologist and in addition by an internist and neurologist with the exception of two cases, who were examined at home by a doctor with neurologic and geriatric experience. Blood tests comprised a minimum of routine hematology and biochemistry, including thyroid functions, vitamins B1, B6, B12 and folic acid. When a CT scan of the cerebrum or an EEG had been performed the results were abstracted from the medical file. The effects of discontinuing potential hallucinogenic medication, such as those listed by Asaad¹⁹ and Weller and Wiedemann³⁰, on the hallucinations were evaluated during a 4-week period.

RESULTS

In a 1-year period, 22 patients were referred for the study. Eight patients did not meet the criteria for the Charles Bonnet syndrome: one patient had no complex hallucinations, two did not show insight, three had delusions and two had additional acoustic hallucinations.

The mean age of the 13 women and 1 man who met the criteria for the Charles Bonnet Syndrome was 81.8 years; 12 of these subjects were living alone. Individual demographic data are shown in table 1.

Description of the hallucinations

Data on the duration of the syndrome and the content of the hallucinations are presented in Table 1. None of the patients attributed a personal meaning to the contents of the hallucinations. When they perceived familiar objects they claimed that these were of no particular interest to them. The hallucinations were not stereotyped in the sense of absolute uniformity: even in 3 patients who always observed the same objects, the number, position, size or intrinsic movement of the objects varied. Four of five in-patients had no hallucinations during the period of hospitalization, but the hallucinations returned after discharge. Further characteristics of the hallucinations are shown in Table 2.

Table 1. Demographic data, duration of syndrome, and content of hallucinations in 14 patients with the Charles Bonnet syndrome

Patient No.	Age (yr)	Marital Status	Duration	Content of hallucinations
A	85	widowed	1.5 yrs	Miniature chimney sweepers, pools of water, plants, flowers, decorative balls from a Christmas tree, white mist.
B	91	widowed	2.5 yrs	Knitting woman, giant man, various eyes and faces, little boy, farmer, various dogs, cat, bird, bunches of flowers.
C	79	unmarried	5 yrs	Various streetscenes with diverse buildings, persons, animals and vehicles.
D	78	widowed	4 months	A group of people conducting mass, various persons with a flower on their heads, various faces, animals and plants.
E	82	widowed	7 yrs	Various people, a horseman, a farmer with plough, plants and flowers.
F	73	unmarried	2 yrs	Humanoid creatures covered in pine branches.
G	88	widowed	3 yrs	Various men, some dressed in doctors' coats and military costumes and a woman with a large black woolly hat.
H	86	divorced	10 yrs	A woman dressed in a torn apron, a very large man dressed in striped pyjamas, various animals and a field.
I	95	widowed	20 yrs	Various young and old couples, strolling children and flowers.
J	74	widowed	5 weeks	Large black spiders crawling on the ceiling (5 years ago she had the same experience for a few months).
K	79	widowed	1 yr	Miniature adults, negro children, dolls, teddy bears, family members and two men lying in her bed.
L	89	married	9 months	A group of people in old-fashioned clothing in her garden and the image of a strolling man on her curtains.
M	70	married	2 yrs	Large variety of people, animals, plants, vehicles, buildings and landscapes.
N	80	divorced	4 weeks	A windmill and a chair.

Table 2. Characteristics of visual hallucinations in 14 patients with the Charles Bonnet syndrome.

Characteristic	No of patients	Characteristic	No of patients
<i>Frequency</i>		<i>Insight in unreal nature</i>	
Daily	9	Prompt full insight	9
Weekly	3	Insight after short while	3
Variable	2	Variable insight	2
<i>Duration</i>		<i>Emotional response</i>	
Minutes	8	Positive	0
Hours	4	Negative	4
Days	2	Indifferent	7
<i>Clearness</i>		Variable	3
Normal	12	<i>Acts stopping hallucinations*</i>	
Greater	2	Closing the eyes	14
<i>Color</i>		Approaching object	6
Normal	12	Looking away	2
Black/White	2	Focussing on object	1
<i>Relationship to surroundings</i>		Switching light on	1
Fitting in well	7	Switching light off	1
Randomly projected	3	<i>Voluntary influence on hallucinations</i>	
Variable	4	Able to evoke	0
<i>Intrinsic movement</i>		Able to influence content	0
Present	6	<i>Facilitating circumstances *</i>	
Absent	1	Home environment	13
Variable	8	Being at rest	9
<i>Familiarity of content</i>		Poor lighting	7
Seen in reality	1	Profound darkness	2
Unfamiliar	8	Bright daylight	1
Both	5		

* Patient may score on more than one item

Psychiatric status

MMSE scores and psychiatric comorbidity are shown in table 3. The low MMSE scores of patients A and E were clearly due to organic mental disorder. The scores of patient C and I however were influenced by uncooperative behavior and serious hearing impairment, respectively.

In all patients with a psychiatric diagnosis except patient A, the onset of the Charles Bonnet syndrome and the mental disorder did not coincide.

Table 3. MMSE scores, psychiatric comorbidity, visual acuity, and presence of abnormal CT cerebrum and EEG in 14 patients with the Charles Bonnet syndrome.

Patient no. EEG**	MMSE score*	concomitant psychiatric diagnoses	visual acuity		CT**	EEG**
			R	L		
A	18/27	organic amnesic syndrome	<0.1	<0.1	+	n.p.
B	25/29	dysthymic disorder	<0.1	0.5	n.p.	n.p.
C	10/25	dysthymic disorder	0.0	0.0	n.p.	n.p.
D	26/30	-	0.5	0.5	+	+
E	13/28	dementia	<0.1	0.25	n.p.	n.p.
F	29/30	-	0.5	0.25	+	+
G	25/30	major depression, recurrent	0.5	0.1	+	+
H	25/30	-	<0.1	0.5	+	-
I	22/27	-	<0.1	<0.1	n.p.	n.p.
J	28/30	major depression, recurrent	1.0	1.0	n.p.	n.p.
K	27/30	-	0.25	<0.1	+	+
L	25/30	-	<0.1	0.25	-	+
M	26/27	-	<0.1	0.0	+	-
N	25/28	-	<0.1	<0.1	+	n.p.

* MMSE score: actual score/highest score possible (owing to visual impairment, many patients could not answer all questions or perform all tasks).

** + = abnormal; - = normal; n.p. = not performed

Medical status

All patients except J were visually impaired (see Table 3). Most of them had more than one eye disease. The diagnoses were: untreated cataract (6 patients), macular degeneration (5), post-cataract extraction (4), retinal ablation (3), glaucoma (3), myopic change of the fundus (1), occlusion of the central retinal vein (1), and optic nerve atrophy (1).

Twelve patients had clinical signs of CNS-disorder, as follows: pyramidal tract signs due to stroke (7 patients), Parkinson's disease (2), oro-facial dyskinesia due to neuroleptic treatment (2), isolated tremor (2) and dyspraxia and dyscalculia of unknown cause (1). The onset of neurological symptoms and hallucinations did not coincide in any of these cases.

The treating physicians had decided not to perform a computed tomography scan in 5 patients and an EEG in 7 patients because the clinical benefit of the results was considered not to outweigh the burden to the patients (see table 3). In the others tomographic findings were as follows: diffuse atrophic changes (8 patients) and multiple cerebral infarcts (3 patients). Electroencephalogram abnormalities consisted of

generalized slowing (5 patients) and various focal disorders (4 patients). No epileptic activity was observed.

In patient D a previously undiagnosed vitamin B 12 deficiency was found; however, adequate supplementation did not lead to a reduction of hallucinations. Blood glucose levels in 5 patients treated for diabetes mellitus and the blood calcium level in 1 patient treated for hypoparathyroidism were adequate.

Only in patient J did we find a possible relationship between medication use and the onset of visual hallucinations. The hallucinations manifested themselves six weeks after she had started taking amitriptyline and disappeared three weeks after the medication was discontinued. A relationship could not be excluded in 2 cases: patient C refused to stop using Timolol eye drops, and in patient N Parkinson's disease was too severe to withhold amantadine and levodopa/benserazide.

Prior to the study, 8 patients had been treated with neuroleptics without any effect on the hallucinations.

DISCUSSION

With the exception of the criterion "hallucinations are stereotyped", the criteria proposed by Gold and Rabins for the Charles Bonnet syndrome were applicable to our group of patients. It is striking that we found no stereotype hallucinations in our patients. They described an often highly variable content and multiformity of the hallucinations. We therefore recommend against including a stereotype nature of the hallucinations as an obligatory characteristic of the Charles Bonnet syndrome.

Two other characteristics were identified that were present in all patients. First, none of the patients showed a tendency towards attributing a personal meaning to the contents of their hallucinations. In this way, these hallucinations distinguished themselves from dream images and from those observed within the framework of schizophrenia or depression. Second, the hallucinations disappeared in all of our patients when they closed their eyes. In the literature, the effect of eye closure is reported to be variable.^{10,22} The other characteristics investigated were variable, as are the reports on these in the literature. In some papers a positive emotional response to the hallucinations is emphasized.^{10,19,22,32} The patients in our population rarely showed a positive response. Some were reluctant to talk about their experiences out of fear that other people, including doctors, would judge them to be mad. The positive emotional experiences described in the literature are probably overrated, because it is likely that patients with such a positive response tend to enjoy sharing their experiences with others. There are more reports on patients who show reluctance.^{9,12,33,34}

In nearly all publications, the authors emphatically distinguish the Charles Bonnet syndrome from the well-known psychiatric syndromes; our study supports this view. We found no relationship of functional psychiatric disturbances to the syndrome. Most authors also state that the cognitive functions are intact, without having any neuropsychologic data available.^{10,31} Others report slight cognitive limitations. In^{22,35} the

majority of our patients we found no evidence of cognitive disturbances on the basis of the MMSE.

Our finding in 8 patients that neuroleptics did not suppress the hallucinations, is in line with the few reports on this subject in the literature.^{25,36} Systematic research has not yet been conducted.

The results of our study support the hypothesis proposed by Gold and Rabins that the Charles Bonnet syndrome is usually associated with ocular pathology and/or cerebral disturbances. Many authors have indicated an association with eye disease.^{9,10,12,22,30,36} However, patient J underlines De Morsier's opinion that the presence of ocular pathology is not obligatory.³¹ The same applies to the presence of CNS-disorder: nearly all patients had signs of CNS-disorder at the time of our study, but we found no clear relationship in time between the onset of neurologic symptoms and hallucinations. In addition, it is unclear whether the atrophic cerebral changes found on the CT scans of these old patients are an expression of disease.

The vast majority of our patients were "single" and living alone. In the GMSS interview, 11 patients indicated that they felt lonely. Without a control group we cannot determine whether social isolation is associated with the syndrome. However, the absence of hallucinations during hospitalization in 4 patients provides some support for a possible aetiological role of social isolation. Hosty made the same observation in two patients.²⁵

Perhaps it is rather the combination of factors that eventually leads to the manifestation of the Charles Bonnet syndrome. Visual impairment, cerebral disorder (structural or functional), and social isolation could be such contributing factors.

Many questions regarding the Charles Bonnet syndrome are still unanswered. It was relatively easy to gather the group of 14 patients we have described here. We therefore think it will be possible to perform an epidemiologic study of this intriguing syndrome in the near future. It is our hope that a better understanding of isolated visual hallucinations will lead us to a more profound insight into the development of this important symptom in other conditions.

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CHAPTER 4

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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ABSTRACT

The aims of this study were to determine the prevalence of the Charles Bonnet syndrome in low-vision patients and analyse possible associated ophthalmic and sociodemographic factors. 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 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. 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. The findings support associations of CBS with sensory deprivation and advanced age.

INTRODUCTION

Since Charles Bonnet in 1760 described complex visual hallucinations in his psychologically normal grandfather, many case reports of "isolated visual hallucinations" have been published.¹ However, systematic research into this phenomenon, which De Morsier called the "Charles Bonnet Syndrome" (CBS), has been limited.² This is probably due to the vagueness of diagnostic criteria and the assumed rarity of the syndrome. Cause and pathogenesis are unknown. Casuistic data suggest associations of the syndrome with old age, eye disease, the female gender, social isolation, bereavement and cerebral disturbance.³⁻⁵ In a recent study of a group of patients with suspected isolated complex visual hallucinations it proved to be feasible to isolate the 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; and absence of delusions and hallucinations in other modalities.⁶

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

PATIENTS AND METHODS

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

The first group ('low vision group') comprised patients from the low vision unit, where visually handicapped patients are referred in order 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 year 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, gender, marital status and social circumstances were obtained from the patients; ophthalmic diagnoses and actual visual acuity status from the ophthalmic records.¹

¹ Visual acuity was measured in each eye separately using 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 on which the patient is still able to discriminate the letters and the distance on which a person with normal eye sight is capable of doing so. Example 1: a patient is able to read letters from a distance of 5 meters 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 discriminate

All the patients underwent a self-designed, semistructured interview by one of three trained interviewers who decided if 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 checklist on hallucinations, the Geriatric Mental State Schedule (GMSS) and the Mini Mental State Examination (MMSE). The psychiatrist decided whether the DSM-III-R criteria for hallucinations and the criteria for the 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; and (b) the period between the first and the last hallucination exceeded four weeks.

In the 'low vision group', we analysed the possible associations between the 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' and the 'optometry group' to investigate whether or not the CBS was directly associated with low visual acuity.

In the statistical analyses the chi-square test was used.

A copy of the screening instrument can be obtained from the authors.

RESULTS

In contrast to only 2 patients (1%) in the 'optometry 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 4 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 2 months to 5 years. In 8 patients, the time criterion was the only reason for not including them as cases. In the remaining 7 patients, the hallucinations had been due to delirium.

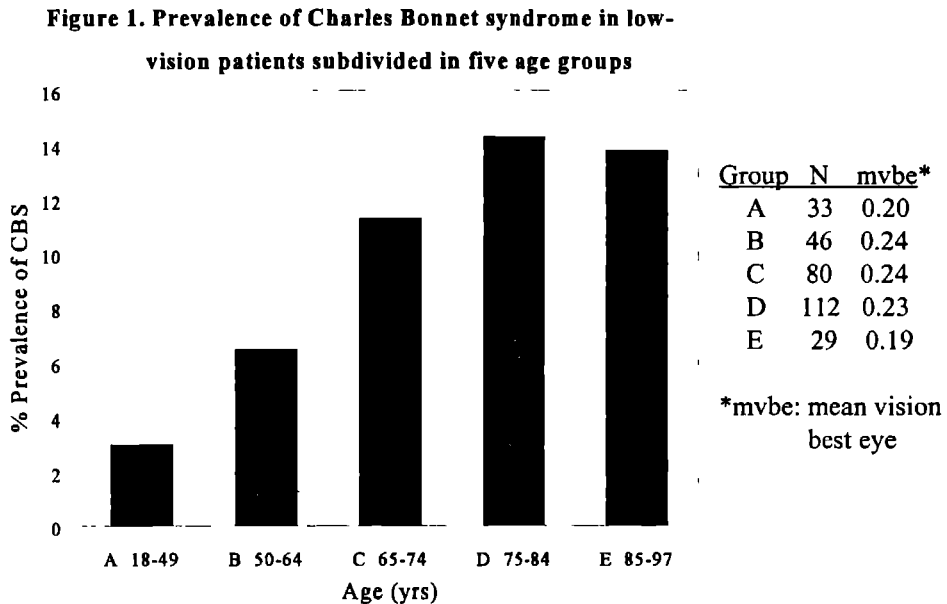
In the 35 cases, the CBS had started from 6 months to 10 years previously. There was a wide variation in the content of the hallucinations: people, animals, flowers and plants, vehicles, buildings, other objects and sometimes complete scenes. Only two patients perceived just a single form. The other patients reported at least 3 different forms.

Apart from a dysthymic disorder in 3 patients no psychiatric disorders were found. The scores on the MMSE ranged from 22 to 30 (mean: 26.5). We considered these

letters from a distance of 5 meters which normally could be read from a distance of 25 meters; visual acuity is expressed as 5/25 or 0.2.

scores to be fair, taking into account the negative influence of visual impairment on the performance of many patients. As before it proved to be feasible to isolate the CBS from other (psychiatric) disorders by the criteria mentioned above.⁶

The prevalence of the CBS in 5 age ranges in the low vision group is illustrated in Figure 1.



The prevalences of the 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, the influence of sociodemographic variables on the prevalence of the CBS was analysed 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 of the 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.

Table 1. Prevalence of the Charles Bonnet Syndrome (CBS) in samples of patients attending the low vision unit ('low vision group') and the optometry unit ('optometry group') of the Department of Ophthalmology in Nijmegen.

Population	N	mean age in yrs.(sd)	vbe* (sd)	% CBS	Statistics on raw data
low vision group	300	70.1 (14.2)	0.23 (0.17)	11.0%	
<i>Age</i>					
18-64 yrs.	79	50.8 (11.7)	0.22 (0.16)	5.1%	$\chi^2=3.861$
> 64 yrs.	221	77.1 (6.5)	0.23 (0.17)	13.1%	$p<0.05$
<i>Ophthalmic diagnoses@</i>					
armd.**	155	76.4 (8.0)	0.22 (0.17)	11.6%	
cataract	125	74.2 (10.0)	0.24 (0.18)	14.4%	
diabetic retinopathy	56	67.4 (10.2)	0.26 (0.14)	16.1%	
glaucoma	37	73.2 (11.2)	0.23 (0.15)	13.5%	
corneal disease	29	76.2 (11.2)	0.26 (0.20)	13.8%	
low vision group >64 yrs.					
<i>Sociodemography</i>					
female	147	77.4 (6.4)	0.23 (0.17)	15.0%	$\chi^2=1.309$
male	74	76.5 (6.7)	0.22 (0.16)	9.5%	$p>0.2$
living alone	97	78.6 (5.8)	0.22 (0.17)	16.4%	$\chi^2=1.725$
not alone	124	75.8 (6.8)	0.23 (0.17)	10.5%	$0.2>p>0.1$
widowed	93	78.8 (6.2)	0.21 (0.16)	17.2%	$\chi^2=2.347$
not widowed	128	75.8 (6.5)	0.24 (0.18)	10.2%	$0.2>p>0.1$
Optometry group	200	74.3 (6.5)	0.73 (0.25)	1.0%	$\chi^2=22.617$
Low vision group >64 yrs.	221	77.1 (6.5)	0.23 (0.17)	13.1%	$p<0.001$
optometry + low vision group >64 yrs. combined					
<i>Visual acuity best eye</i>					
0.01-0.30	180	76.9 (6.6)		14.4%	$\chi^2=23.114$
> 0.30	241	74.9 (6.2)		2.1%	$p<0.001$
0.01-0.15	99	77.3 (6.6)		13.1%	
0.16-0.30	81	76.5 (6.6)		16.0%	
0.31-0.60	104	77.3 (6.2)		2.9%	
0.61-1.25	137	73.1 (6.3)		1.5%	

* vbe = visual acuity in best eye

** armd = age-related macular degeneration

@ Only diagnoses present in more than 20 patients are given. Many patients had more than one diagnosis. The diagnoses listed were not necessarily the major cause of visual impairment. Cataract was never the major cause (patients with severe cataract would have been operated on instead of being referred to the low vision unit).

DISCUSSION

In the literature, we found only 2 prospective case-finding studies on the CBS. Olbrich et al detected 5 cases of the CBS in 43 elderly low vision patients, whereas Berrios and Brook found 2 cases in 150 psychogeriatric patients.⁷⁻⁸ In other prevalence studies on complex visual hallucinations in visually impaired patients the concept of the CBS was not used and no general psychiatric examination was conducted.

Our study was the first major case-finding study on the CBS in low vision patients and the only one which used fixed criteria and a control group. The prevalence of the CBS in our low vision population was comparable with the findings of Olbrich et al, 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 and Brook.⁷⁻⁸

The 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 the 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.⁷ Many of our patients were quite relieved to hear that the 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 the 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 the CBS, as was demonstrated by our finding of only one case in 60 of such patients. One other study reported an association between complex visual hallucinations (although not defined as the CBS) and bilateral impaired vision.⁹

Our finding that the CBS is associated with advanced age supports similar assumptions in the literature.²

The prevalence of the CBS differed little between 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 the women clearly outnumbered the men.^{4,6} In this study we found the prevalence of the CBS to be higher in women than in men, but we could not statistically confirm a sex effect.

The CBS is thought to be related to social isolation.⁶ Holroyd et al found a significant association between complex visual hallucinations and the variable "living alone" in their ophthalmic population.⁹ In our study (on a larger population and restricted to the CBS) we found a trend in this direction, but could not confirm the hypothesis statistically. The same applies to the possible association between the CBS and bereavement.

We conclude that the CBS is associated with low visual acuity and advanced age, as two independent factors.

The association between the CBS and visual impairment supports those theories on the pathogenesis of the syndrome in which sensory deprivation has a role.⁵ A reduction of visual input, however, cannot be the sole cause of the CBS, because not all visually handicapped have hallucinations, and the CBS has been described in patients with normal vision.¹ 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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CHAPTER 5

Visual hallucinations in psychologically normal people: Charles Bonnet's syndrome.

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ABSTRACT

The Charles Bonnet Syndrome (CBS), characterised by the presence of complex visual hallucinations in psychologically normal people, was considered for a long time to be rare. Systematic research on CBS has been limited. However, it has been realised that CBS occurs frequently in elderly, visually handicapped patients, and we have been able to study the syndrome in a large number of patients. After screening 505 visually handicapped patients, 60 were found to meet proposed diagnostic criteria for CBS. Psychopathological characteristics, personal meaning, and the emotional impact of hallucinations, as well as factors influencing the hallucinations, were analysed. Although diagnostic criteria demand merely "partial insight", all patients had full insight into the unreal nature of their hallucinations. Other characteristics varied. In 46 (77%) patients, hallucinations lacked a personal meaning. Sensory deprivation and a low level of arousal seemed to favour the occurrence of hallucinations. CBS caused considerable distress in only 17 (28%) patients. However, all patients were glad to be told that their hallucinations were not due to mental disease. The proper diagnosis been made in only one of the 16 patients who had consulted a doctor.

INTRODUCTION

The Charles Bonnet Syndrome (CBS), named after the Swiss philosopher who described the phenomenon for the first time in 1760, is characterised by the presence of complex visual hallucinations in psychologically normal people.^{1,2} For a long time the syndrome was considered to be rare. In 1989 Podoll and coworkers counted only 46 patients who had been described in the literature since 1760.³ Probably due to the presumed rarity of the condition and the lack of generally accepted diagnostic criteria, systematic research on the CBS has been limited. The present state of knowledge on the CBS, based on single case reports and some small case series, may be summarized as follows: CBS patients have complex visual hallucinations which cannot be explained by the presence of a psychiatric disorder. They have insight into the unreal nature of their perceptions. The course of the syndrome may be episodic, periodic or chronic. Response to neuroleptic medication is poor. The majority of patients are old and already have disorders of the eyes or central visual system. Many questions concerning the clinical characteristics of the syndrome, its etiology and pathogenesis remain unanswered.²⁻⁷

Several authors have questioned the rarity of the CBS. They suggested that many patients did not report their hallucinations out of fear of being considered insane.⁸⁻¹¹ Diagnostic criteria for the CBS have been proposed.^{5,11} Associations of the CBS with advanced age and low visual acuity were demonstrated and a prevalence of 11% in a group of 300 visually handicapped patients was found.¹²

In this article, the results of a further study on 60 patients with CBS are given. Our aims were to explore the psychopathological characteristics of the hallucinations, their personal meanings and influencing factors and to assess the extent of distress associated with the syndrome.

PATIENTS AND METHODS

A case-finding study was undertaken at the Low Vision Unit of the Department of Ophthalmology, University Hospital Nijmegen, to which visually handicapped patients are referred to receive reading aids from an optometrist. Of 511 consecutive patients older than 18 years, 505 gave their informed consent to participate in the study.

Data were collected on demographic characteristics, ophthalmic diagnoses, and visual acuity. All patients underwent a semistructured interview on complex visual hallucinations with one of four trained interviewers. In cases of possible or probable complex visual hallucinations, the patient's informed consent to a further investigation was asked. This investigation comprised an interview by a psychiatrist (RJT) at the patient's home. The psychiatrist checked whether the definition of hallucinations of the Diagnostic and Statistical Manual of Mental Disorders was met: "A sensory

perception without external stimulation of the relevant sensory organ.”¹³ A general psychiatric examination was performed with the Dutch language version of the Geriatric Mental State Schedule.¹⁴⁻¹⁵ The psychiatrist then decided whether the following criteria for the CBS were met: 1) at least one complex visual hallucination within the past four weeks; 2) a period between the first and the last hallucination exceeding 4 weeks; 3) full or partial retention of insight into the unreal nature of the hallucinations; 4) absence of hallucinations in other sensory modalities; 5) absence of delusions. Using a checklist, the patients were interviewed with special attention to psychopathological characteristics, personal meaning of the hallucinations, factors encouraging or stopping the hallucinations and the emotional impact of the hallucinations on the patient.

RESULTS

Sixty-three of the 505 visually handicapped patients had experienced complex visual hallucinations in the 4-week period before screening; one patient did not meet CBS criteria because of lack of insight and coexistence of acoustic hallucinations; two patients refused further examinations.

Eighteen men and 42 women with the CBS remained. Their ages ranged from 46 to 98 years with a mean age of 75.4 (sd 8.0) years. Mean visual acuity in the best eye was 0.23 (sd 0.18). The most frequent causes of visual impairment were age-related macular degeneration (31 patients), diabetic retinopathy (11 patients), glaucoma (4 patients) and corneal disease (3 patients). Eleven patients had less common ophthalmic diagnoses. The psychiatric examination of the patients revealed no disorders, which could be considered to be the cause of their hallucinations.

The mean age of onset of the CBS was 72 (sd: 5) years. The duration of the syndrome at the time of screening ranged from 1 month to 30 years: 29 patients had experienced hallucinations for less than a year, 21 between 1 to 5 years and 10 for more than 5 years.

Psychopathological characteristics of the hallucinations

The frequency of hallucinatory episodes varied from several times daily to only twice a year. In the course of time the frequency had decreased in 10 patients, had increased in 2 patients and had remained unchanged in 48 patients. Hallucinatory episodes had lasted from a few seconds to many hours. The patients described the content of their hallucinations as people, animals, plants, a large variety of inanimate objects and sometimes complete scenes. Often, the content of hallucinations was mundane (an unfamiliar person, a bottle, a hat), but it could be funny (2 miniature policemen guiding a midget villain to a tiny prison van), ghostly (translucent figures floating in the hallway), bizarre (a dragon, people wearing one big flower on their heads), as well as beautiful (a shining angel, wonderful bunches of flowers). Most patients described a large variety of hallucinations, differing in each hallucinatory episode. Sometimes the sensation of specific objects returned, but stereotyped hallucinations (identical in every respect) were uncommon. Hallucinations contained both familiar and unfamiliar images. The hallucinations occurred both in black and white or colour. They could be clearer, equally clear or less clear in comparison to reality. They could show intrinsic movement, a movement of the total image, or be motionless. Sometimes they moved along with the eyes. Most patients hallucinated only with their eyes open. Some perceived hallucinated objects as floating in the air or projected on a wall or ceiling. Others reported that the objects fitted well into the surroundings (e.g. an unreal person sitting in a real chair). Patients hallucinating while their eyes were closed perceived hallucinations in the dark subjective space in front of the eyes. The numbers of patients who reported these characteristics are listed in Table 1.

Table 1: Characteristics of hallucinations in 60 patients with Charles Bonnet syndrome

Characteristic	n (%)	Characteristic	n (%)
Frequency*		Duration	
daily	16 (27)	1-5 seconds	8 (13)
weekly	18 (30)	5-60 seconds	10 (17)
monthly	21 (35)	1-60 minutes	32 (53)
less often	5 (8)	> 1 hour	9 (15)
		not certain	1 (2)
Content†		Familiarity of content	
people	48 (80)	always	5 (8)
-adults	42 (70)	sometimes	15 (25)
-faces	21 (35)	never	39 (65)
-children	14 (23)	not certain	1 (2)
-large groups	7 (12)		
-miniature of people	2 (3)	Influence eyelids	
animals	23 (38)	only with opened eyes	40 (67)
plants/trees	15 (25)	only with closed eyes	8 (13)
buildings	9 (15)	variable	12 (20)
scenes	9 (15)		
other objects	25 (42)	Moving along with eyes	
Relationship to the surroundings‡		always	11 (18)
fitting in well	13 (22)	sometimes	9 (15)
randomly projected	22 (37)	never	32 (54)
variable	17 (28)	not certain	8 (13)
Presence of colours		Clarity	
always	38 (63)	as real objects	16 (27)
sometimes	6 (10)	clearer	27 (45)
only black/white	16 (27)	less clear	12 (20)
		variable	5 (8)
Intrinsic movement		Movement "en bloc"	
always	18 (30)	always	13 (22)
sometimes	10 (17)	sometimes	2 (3)
never	32 (53)	never	43 (72)
		not certain	2 (3)
Recurring objects		Stereotyped hallucinations	
always	13 (22)	always	6 (10)
sometimes	11 (18)	sometimes	3 (5)
never	36 (60)	never	51 (85)

* Mean frequency over last 3 months is given. In 19 patients the frequency was variable: days/weeks with many hallucinations were followed by weeks with no or few hallucinations.

† More than 1 item may be mentioned.

‡ Only applicable to patients hallucinating with opened eyes.

Forty-nine patients (82%) stated they were always immediately aware of the unreal nature of their hallucinations. Eleven (18%) patients had sometimes been deceived for a short period, but this had happened only when hallucinated objects looked 'ordinary' and fitted realistically in the surroundings. The patients were always easily corrected by others while hallucinating: a lady told us she once sat at her window watching cows in a neighbouring meadow. It was actually very cold and in the middle of winter, and she complained to her maid about the cruelty of the farmer. The astonished maid, however, saw no cows. Embarrassed, the patient then admitted that her eyes were no longer to be trusted.

Personal meaning of the hallucinations

Forty-six (77%) patients could not detect any personal relevance of the hallucinations. Even if hallucinated objects were familiar to them, they were emotionally of no apparent importance. One woman compared her hallucinations with her dreams: "In my dreams I experience things which affect me, which are related to my life. These hallucinations however have nothing to do with me."

Three (5%) patients were uncertain whether or not some of their hallucinations had a personal meaning: an elderly childless gentleman was intrigued by recurrent hallucinations of a little girl and boy. He wondered whether these hallucinations reflected his unfulfilled wish to become a father.

Ten (17%) patients experienced hallucinations involving emotionally important as well as unimportant objects. For instance, one patient perceived a great number of people, most of whom were unfamiliar, but occasionally a deceased relative appeared among them.

Only one (2%) patient had hallucinations with an exclusively personal emotional relevant content: these always involved her late husband. Since his death 3 years earlier she experienced these hallucinations several times weekly.

Factors influencing hallucinations

No patient was able to consciously evoke hallucinations or exert influence on their content. Many, however, had noted circumstances which seemed to favour the occurrence of hallucinations and/or had found methods to stop hallucinating. Details are shown in Table 2.

Emotional impact of the hallucinations

The emotional response to the hallucinations was mainly negative in 19 (32%) patients: anxiety in 14 and irritation in 5 patients. Eleven (18%) patients showed mixed emotions and in 22 (37%) patients the emotional response was neutral. Eight (13%) patients had felt joy or amusement in their experiences. General feelings of well-being were not disturbed by the hallucinations in 43 (72%) patients. Seventeen (28%) patients suffered from their hallucinations and hoped that they would disappear; only six felt enough distress to consider taking medication to suppress their hallucinations.

Table 2: Circumstances favouring and acts stopping hallucinations in 60 patients with the Charles Bonnet syndrome (CBS).

Favourable circumstances studied in all patients	n (%)	Acts stopping hallucinations studied in all patients	n (%)
Specific times of day		Influence eyelids*	
-evening	21 (35)	-keeping eyes closed	23 (38)
-night	14 (23)	-keeping eyes opened	9 (15)
-midst of day	7 (12)	Approaching hallucinated object*	12 (20)
-early morning	6 (10)		
Light intensity			
-poor lighthing	39 (65)		
-bright daylight	9 (15)		
Being inactive	51 (85)		
Home environment	43 (72)		
Being alone	23 (38)		
Other favourable circumstances†	n (%)	Other acts stopping hallucinations†	n (%)
Fatigue	6 (10)	Looking/walking away	9 (15)
Nervousness/stress	5 (8)	Putting on a light	6 (10)
Watching television	3 (5)	Blinking	5 (8)
Sitting in a car	3 (5)	Moving eyes swiftly	3 (5)
Laser therapy eyes	1 (2)	Fixing hallucination with eyes	3 (5)
Using Temazepam	1 (2)	Concentrating on something else	3 (5)
		Hitting hallucination	1 (2)
		Shouting	1 (2)

* The effect of these acts had not been tried out by all patients.

† Answers to the question "have you noticed other favourable circumstances and/or methods to stop hallucinations?".

Forty-four (73%) patients had not mentioned their extraordinary experiences to doctors: 15 patients feared their doctor would not take them seriously or would think they were insane; 20 patients thought this was not the kind of complaint for which one consults a doctor; 9 patients gave no explanation. Only 1 of the 16 patients who had consulted a doctor (mostly a general practitioner or ophthalmologist) was informed about the proper diagnosis. Seven patients had experienced the doctors' reaction as

negative. A patient who consulted a psychiatrist because of family problems and then mentioned her hallucinations promptly received neuroleptic treatment. This treatment had not suppressed hallucinations but had only made her feel awkward. The general practitioner of another patient had responded with: "you'd better not talk about such silly things!"

DISCUSSION

The clinical characteristics of the CBS showed rich variety. We found no characteristics which were common to all patients in addition to the inclusion criteria we used. However, in this population the criterion "full or partial insight" could be sharpened to "full insight". Some patients had occasionally needed correction by others, but this was because the ordinary appearance of their hallucinations, fitting well in the surroundings, made it very difficult or even impossible to discriminate real from unreal.

There is a continuing discussion in the literature as to whether the CBS-type hallucinations are newly created products of fantasy or reproductions of earlier true perceptions.^{3,4,6} The fact, that the majority of hallucinations of our patients contained objects that they did not remember having seen in reality seems to support the first theory. However, sometimes familiar objects were recognized, though one could argue that these familiar objects are also new creations, which are modeled after earlier true perceptions. In favour of the second theory it could be suggested that the patients had once seen the unfamiliar objects in reality, but had forgotten them because they were of no particular interest. We can not bring this discussion to a conclusion.

Some authors have suggested that the content of the hallucinations in CBS is influenced by wishes or preoccupations of the patients.^{4,7} This theory seemed probable in only a minority of the patients in our study. The infrequent occurrence of emotionally important objects in the large variety of hallucinations may just be coincidental.

The circumstances favouring hallucinations provide some support for the suggestion that sensory deprivation and a low level of arousal are triggers for CBS type-hallucinations.^{3,4,6,10,16} Circumstances such as "early morning", "evening" and "night", "being inactive", "home environment", "fatigue" and "using temazepam" might be associated with a low level of arousal. "Poor lighthouse", "being alone" and "laser therapy for the eyes" may point to sensory deprivation. Possibly typical acts that stop hallucinations reflect an increase in the level of arousal and/or sensory stimulation.

The CBS had little impact on the general feelings of well-being in the majority of patients. Nevertheless, all of them were glad to be informed that it was a known phenomenon, which had a name and was not considered to be a mental disorder. This study shows that many patients do not consult a doctor about the CBS. We found that

patients tended to conceal their extraordinary experiences from others as well: “Why have you not told me about this?”, the amazed wife of one man asked as he confessed to the interviewer that he perceived “faces” since 3 years. “I didn’t want to upset you”, he replied.

The experiences of those patients who sought professional advice also indicate that many doctors are not familiar with the CBS. Patients would benefit if doctors recognized the CBS and gave them proper information, including reassurance that they are not mentally ill. For most patients this response will be sufficient. Support should be offered to those who cannot cope with their hallucinations. No treatment of proven effectiveness is yet available.

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CHAPTER 6

Somatic and psychiatric characteristics of patients with the Charles Bonnet syndrome: a controlled study

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ABSTRACT

The Charles Bonnet syndrome (CBS) is characterized by the presence of complex visual hallucinations in non-psychotic people. Recognized risk indicators for CBS are advanced age and impaired vision. In this study possible other risk indicators were examined in elderly patients from a low vision unit. Using a case control approach 52 patients with CBS and 80 control patients were interviewed on medical history, current medical complaints, medication, and hearing loss and examined with Mini-Mental State Examination and Geriatric Mental State Schedule. CBS was significantly associated with energy loss, a larger number of comorbid somatic disorders and β -blocking medications. No evidence was found for associations of CBS with cerebral and metabolic disorders, hearing loss, cognitive status, and psychiatric disorders. The findings suggest that energy loss, connected with compromised general health, and β -blocking medication favour the occurrence of CBS in visually handicapped elderly.

INTRODUCTION

In 1760 the Swiss philosopher Charles Bonnet described the vivid complex visual hallucinations of his psychologically normal grandfather. Since then the phenomenon of 'isolated visual hallucinations' -later to be known as the 'Charles Bonnet syndrome' (CBS) has been repeatedly observed, often in elderly ophthalmic patients. The clinical characteristics of the syndrome are quite variable, but typically, in most patients reality-testing remains intact. Little is known about etiology of CBS.¹⁻² Recently, associations of CBS with advanced age and low visual acuity were statistically confirmed.³

In theories on the pathogenesis of CBS the role of visual impairment is often stressed. However, only a minority of the visually handicapped experience hallucinations and CBS may occur in persons with intact vision. Other factors have to be involved. Based mainly on casuistic data, many aetiological and risk factors have been postulated, including cerebral disorders, metabolic disorders, side-effects of medication, impaired general health and cognitive impairment.¹⁻⁵ The aim of the present case control study, involving the population at risk of elderly visually handicapped patients, was to find statistical evidence of CBS connections to somatic and psychiatric factors.

PATIENTS AND METHODS

A case-finding study was undertaken at the Low Vision Unit of the Department of Ophthalmology, University Hospital Nijmegen, to which visually handicapped patients are referred to receive reading aids from an optometrist. Consecutively referred patients older than 64 years underwent a semistructured interview on complex visual hallucinations. Possible cases were examined by a psychiatrist who investigated whether the following criteria for CBS were met: a) presence of complex, persistent, or repetitive visual hallucinations; b) full or partial retention of insight; c) absence of delusions; d) absence of hallucinations in other sensory modalities.^{4,6} Of 373 patients 52 (14%) had CBS. We reported on the characteristics of CBS in these patients, along with 8 younger patients, elsewhere.⁷

All 52 CBS patients and 80 out of 97 consecutive patients without complex visual hallucinations gave their informed consent to participate in this study. By using a checklist, patients were interviewed on demographic data, medical history, current medical complaints, medication and hearing problems. Cognition was examined with the Mini Mental State Examination (MMSE) and psychiatric status with the Geriatric Mental State Schedule (GMSS).⁸⁻¹⁰ Ophthalmic data were abstracted from the ophthalmic charts.

Data were analysed by using the SPSS software package. Regarding categorical variables CBS patients and controls were statistically compared by chi-square tests. For the comparison of continuous variables t-tests were applied. In the analysis of psychiatric symptoms, the large number of GMSS items necessitated variable reduction. This was achieved by excluding a) items not referring to psychiatric symptoms; b) items referring to in- and exclusion criteria for CBS and controls (hallucinations and delusions); c) cognitive test items (already analysed using MMSE); and d) items with positive scores in less than 10% of the study-population (CBS and control patients combined). The remaining 65 dichotomous variables were combined, using factor analysis. Reliability of these factors was studied by Cronbach's α . Logistic regression and the forward variables selection procedure, with application of Bonferroni's correction for statistical significance, was then used to analyse associations with CBS.

RESULTS

Mean age was 78.4 (sd 5.7) years in the 52 CBS patients and 77.2 (sd 6.3) years in the 80 control patients ($t=1.11$, $p=0.27$). Sixty-nine percent of CBS and 60% of control patients were female ($\chi^2=1.16$, $p=0.28$). Mean visual acuity of the better eye was 0.21 (sd 0.17) in CBS and 0.23 (sd 0.18) in the control patients ($t=0.76$, $p=0.46$). Most frequent causes of visual handicap were age-related macular degeneration (60% of CBS subjects and 70% of controls; $\chi^2=1.51$, $p=0.22$) and diabetic retinopathy (13%

of CBS subjects and 9% of controls; $\chi^2=0.74$, $p=0.39$).

In Table 1 the main results regarding somatic comorbidity, current medication, hearing loss and cognitive status are given. To the table we may add that no significant associations were found between CBS and the most frequent specific cerebral and metabolic disorders: (post) stroke (12% in CBS, 13% in control patients) and diabetes mellitus (29% in CBS and 20% in control patients). Furthermore, there were no indications for an epileptic or migrainous cause of hallucinations.

Table 1: Somatic comorbidity, medications, hearing loss and cognitive status in 52 patients with Charles Bonnet syndrome and 80 control patients

	CBS patients		Control patients		Statistics	
	%	Mean (sd)	%	Mean (sd)	Test value	P
Somatic comorbidity *						
Number of disorders		3.3 (2.0)		2.3 (1.6)	t=3.23	0.002
Prevalence of disorders ⁺						
-Cardiovascular	77		61		$\chi^2=3.52$	0.06
-Metabolic	42		30		$\chi^2=2.10$	0.15
-Musculo-skeletal	39		26		$\chi^2=2.19$	0.14
-Cerebral	19		23		$\chi^2=0.20$	0.65
-Pulmonary	17		11		$\chi^2=0.98$	0.32
-Gastro-intestinal	14		14		$\chi^2=0.00$	0.96
Current medication						
Number of medications		3.8 (2.6)		3.1 (2.4)	t=1.68	0.09
Proportion using: ⁺						
-anticholinergics [#]	56		46		$\chi^2=1.14$	0.29
-benzodiazepines	35		25		$\chi^2=1.42$	0.23
- β -blocking agents	38		18		$\chi^2=7.24$	0.007
-thiazide diuretics	21		19		$\chi^2=0.12$	0.73
-calcium antagonists	21		15		$\chi^2=0.83$	0.36
Hearing loss	65		59		$\chi^2=0.59$	0.44
Mini Mental State Exam.	26.5 (2.0)		26.3 (3.1)		t=0.50	0.63

* Ophthalmic disorders not included.

⁺ Only categories present in >10% of combined groups are listed.

[#] Medications with anticholinergic (side)effects.¹¹

Psychiatric disorders, mostly minor affective- and anxiety disorders, were found in 27% of CBS and 18% of control patients ($\chi^2=1.67$; $p=0.20$). In all cases hallucinations and psychiatric disorder were unrelated.

Factor analysis of the 65 GMSS items resulted in factors, which were difficult to interpret because of (a) the wide variety of symptoms to which they referred and (b) dispersion over several factors of items which were closely related from a clinical point of view. This was probably due to the fact that few patients had fully developed psychiatric syndromes. As the 65 items referred to only a limited number of symptoms, it was decided to combine them into the following 10 item clusters: 'worries', 'anxiety', 'tension', 'depressed mood', 'subjective memory problems', 'sleep disturbances', 'unsteadiness', 'energy loss', 'loneliness', and 'obsessive symptoms'. A few items were found to reduce the reliability of these clusters. Depending on clinical considerations, these were omitted or left in. Cronbach's α of the 10 clusters, finally representing 56 GMSS-items, ranged from 0.67 ('tension') to 0.86 ('obsessive symptoms'). Applying Bonferroni's correction, only p values <0.005 ($0.05/10$) were considered to indicate statistical significance in the following logistic regression analysis. This resulted in 'energy loss' as the only significant risk indicator ($p=0.0001$).

DISCUSSION

Advanced age and low visual acuity are recognized riskfactors for CBS.³ Differences regarding these variables between the present CBS- and control group were insignificant because of the chosen study design.

Energy loss was a highly significant risk indicator for CBS in this population. It is unlikely that CBS itself causes a loss of energy, as CBS is responsible for little or no distress in most patients.⁷ Compromized general health, as indicated by the significantly larger number of comorbid somatic disorders in CBS patients, is a more plausible cause. A lack of energy could be an etiological factor for CBS: CBS patients have mentioned 'fatigue' as a factor encouraging hallucinations.⁷ Lack of energy could contribute to the 'low level of arousal' that triggers CBS-type hallucinations according to some authors.¹ The association between the use of β -blocking agents and CBS probably reflects a causal relationship as well. Hallucinations have been described as rare side-effects of these drugs.¹² Our findings suggest an unusually high risk for this side-effect in visually handicapped elderly.

No connections of CBS with cerebral disorders and/or cognitive impairment were found. However, the importance of this finding is limited, because of the low prevalence of cerebral disorders in the study population. Severe cognitive impairment was rare: only two patients, both controls, had dementia. The negative findings regarding associations between CBS and psychiatric syndromes and symptoms are in line with the concept of CBS as an isolated phenomenon from a psychiatric point of view.

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CHAPTER 7

Social and psychological characteristics of patients with the Charles Bonnet syndrome

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ABSTRACT

The Charles Bonnet syndrome (CBS) is characterized by the presence of complex visual hallucinations in psychologically normal people. Little is known about aetiology and pathogenesis of CBS. Recently, advanced age and low visual acuity proved to be risk indicators. Aim of this study was to find possible psychological and social risk indicators.

All subjects were patients over the age of 64 years from a low vision unit. Using a case control approach, 50 patients detected as having CBS and 80 patients without visual hallucinations were interviewed on their educational level, social circumstances, number of social contacts and ability to cope with their visual handicap. Loneliness was measured with the De Jong-Gierveld loneliness scale and personality traits were examined with the Dutch language short version of the Minnesota Multiphasic Personality Inventory (NVM).

Compared to the control group, significantly more CBS patients were lonely. Mean scores on NVM shyness scale were significantly higher and on NVM extraversion scale significantly lower in CBS patients. In multiple logistic regression analysis for the 3 determinants simultaneously, loneliness and low extraversion were highly significant predictors for CBS, whereas the predictive effect of shyness was not significant.

It is concluded that loneliness, low extraversion and shyness are risk indicators for CBS in elderly visually handicapped people.

INTRODUCTION

In 1760 the Swiss philosopher Charles Bonnet described the occurrence of vivid complex visual hallucinations in his psychologically normal grandfather.¹ Since then, the phenomenon of 'isolated visual hallucinations' -later to be known as the 'Charles Bonnet syndrome' (CBS)- has been repeatedly observed, often in elderly ophthalmic patients.²⁻⁵ Characteristics of hallucinations showed a rich variety, but a common feature of most patients was that their reality testing remained intact. Little is known about aetiology and pathogenesis of CBS. Systematic research has been limited, probably because of the assumed rarity of the syndrome and the lack of generally accepted diagnostic criteria.

Recently the following diagnostic criteria were proposed: a) presence of formed and complex, persistent or repetitive visual hallucinations; b) full or partial retention of insight; c) absence of delusions; d) absence of hallucinations in other sensory modalities.⁶⁻⁷ Using these criteria, significant associations of CBS with advanced age and low visual acuity were demonstrated and a prevalence of 12% in a group of 505 visually handicapped adults was found.⁸⁻⁹

Visual impairment is probably an important aetiological factor in many cases. However, only a minority of the visually handicapped experience visual hallucinations and CBS has been found in persons with intact vision as well.^{2,4} Thus, other factors have to be involved. In the older literature an alleged high incidence of CBS among philosophers, psychologists and university professors led to remarks on developed powers of observation and imaginative memory as predisposing personality characteristics.¹⁰⁻¹¹ In more recent literature predisposing social factors, including bereavement and social isolation are postulated.^{7, 12-14} These assumptions are all based on findings in small case series or even single cases. In the only systematic study on personality Schultz & Melzack found no striking common profiles in 14 CBS patients.¹⁵ In a recent prevalence study CBS was found to occur more frequently in widowed people and people living alone, but associations did not reach statistical significance.⁸ In two studies concerning visual hallucinations 'in general' in the risk group of visually handicapped patients, significant associations with 'maladaptive coping with visual loss' and 'living alone' were found.¹⁶⁻¹⁷ However, the information given in these studies is insufficient to determine how many CBS patients were among the hallucinators.

Aim of the present study was to examine possible psychological and social risk indicators for CBS. Using a case-control model, associations between CBS and level of education, ability to cope with visual handicap, social circumstances, loneliness and personality characteristics were analysed in a population of visually handicapped elderly people.

METHOD

Patients

All subjects were patients from the Low Vision Unit of the Department of Ophthalmology of the University Hospital Nijmegen, to which visually handicapped people are referred to receive reading aids from an optometrist. We reported on a case-finding study in this population and the clinical features of CBS in detected cases elsewhere.⁹ In short: 505 adult patients underwent a semistructured interview on visual hallucinations. Possible cases were examined by a psychiatrist, using the Dutch language version of Geriatric Mental State Schedule, who decided whether the criteria for CBS described above were met.^{6-7, 18-19} Sixty patients with CBS were found.

The 52 patients with CBS, who were over the age of 64 years, were asked to participate in the present study. Fifty gave their informed consent. Eighty out of 97 consecutive patients over the age of 64 years, who had experienced no complex visual hallucinations, gave their informed consent to participate as control patients.

Assessment

All 130 patients underwent a standardized study protocol. Ophthalmic data were abstracted from the ophthalmic charts. Patients were interviewed at home by one of four trained interviewers on the following items:

(a) *Sociodemography*

Information was obtained on marital status, living situation and level of education. The patients were asked how often they had visited, and had been visited by, their children, other relatives and friends/acquaintances in the past four weeks.

(b) *Loneliness*

The rasch scale developed by De Jong-Gierveld & Kamphuis was used. This scale contains 12 statements connected with loneliness. Patients are asked to determine whether the statements are appropriate in their case. Answers indicating absence of loneliness are rated 0 points, neutral answers and answers indicating loneliness are rated 1 point. Persons with a total score of 0-3 are considered to be not lonely, with a score of 4-6 moderately lonely and with a score of 7-12 very lonely.²⁰

(c) *Personality traits*

The 'Nederlandse Verkorte MMPI' (NVM), a Dutch language short version of the Minnesota Multiphasic Personality Inventory (MMPI) was used. Research on this well validated scale has shown that the shortening of the MMPI has not led to a significant loss of information. The NVM contains 83 items spread over 5 scales. Each item contains a statement of which the patient has to decide whether or not it is appropriate in his/her case. If the patient is unable to decide he/she may give a neutral answer. Answers indicative of a specific personality trait are rated 2 points, neutral answers 1 point, and answers not indicative 0 points. The 5 NVM scales are:

1. Negativism (22 items): refers to passive avoidant behavior, feelings of dissatisfaction and grudge regarding daily life events and aggressive behavior. Persons with high scores show a negative, dissatisfied and hostile attitude towards

other people and tend to react with grumbling and complaining to feelings of discomfort. They often feel tense and behave self-defensive. In persons with a low score this is all not the case. This scale is related to the Psychopathic Deviate, Hypomania, Lie, Depression, Masculinity- Femininity and Schizophrenia scales of the MMPI.

2. Somatisation (20 items): contains items connected with vague physical complaints. Persons with high scores tend to react to psychological stress with physical symptoms. Persons with low scores do not. This scale is related to the Hypochondriasis, Hysteria and Depression scales of the MMPI.

3. Shyness (15 items): items reflecting feelings of shyness and difficulties in the interpersonal contacts. People with high scores indicate to feel shy often and to regard this as a problem. They are reserved, introverted, silent, and less sociable. Persons with low scores don't feel shy, are jovial and talkative. This scale is related to the Social Introversion, Hysteria and Psychopathic Deviate scales of the MMPI.

4. Psychopathology (13 items): items connected with delusional feelings, paranoid thoughts and bizarre experiences. These are present in persons with high scores. Low and average scores should be considered normal. In the present study this scale was reduced to 12 items because one item implied the presence of visual hallucinations, which were by definition always present in CBS patients and, as an exclusion criterion, absent in control patients. This scale is related to the Paranoia, F and Schizophrenia scales of the MMPI.

5. Extraversion (13 items): items reflecting an active and energetic attitude towards various social contacts. Persons with high scores are extraverted, persons with low scores are not. This scale provides no indication for introversion. It is related to the Social Introversion and Depression scales of the MMPI.

The authors of the NVM provide 3 norm tables -general population, psychiatric patients, somatic patients- to classify scores in 7 categories ranging from 'very low' to 'very high'. In this study the norm table for somatic patients was used for comparative purposes.²¹⁻²³

(d) *Coping with visual handicap*

Patients were asked 3 questions: (1) 'Can a person cope with an eye disease such as yours?', (2) 'To what extent does your eye disease interfere with your joy in life?', and (3) 'Have you learned to live with your eye disease?' Patients had to choose one of five answers to each question. Answers ranged from minimal coping (rated 0 points) to maximal coping (4 points). Adding up the scores led to a 3 item scale with a total score ranging from 0 (minimal coping) to 12 (maximal coping). Reliability of this scale was determined using Cronbach's alpha.

The De Jong-Gierveld loneliness scale and the NVM are self rating scales. Because of the reading difficulties of these visually handicapped patients, the items were read aloud to the patients by the interviewers. The interviewers were instructed to act just as reading aids and not to influence the patients.

Statistical analysis

Data were analysed using the SPSS software package. Regarding categorical variables CBS patients and controls were statistically compared by chi-square tests. For the comparison of quantitative variables, t-tests were applied. Finally, multiple logistic regression analysis was used to compare the magnitudes of the predictive effects for CBS among the determinants found significant in the univariate analyses.

RESULTS

Ages ranged from 67 to 97 years in the 50 CBS patients and from 66 to 93 years in the 80 control patients. Mean age was 78.4 (sd 5.8) in the CBS and 77.2 (sd 6.3) in the control group. Seventy-two percent of CBS and 60% of control patients were females. Mean visual acuity of the best eye was 0.21 (sd 0.17) in CBS and 0.23 (sd 0.18) in control patients. With regard to these variables CBS and control group were not significantly different: mean age: $t=0.81$, $p=0.31$; gender: $\chi^2=1.94$, $p=0.16$; visual acuity: $t=0.81$, $p=0.42$.

The results of the analyses of the variables education, coping with visual handicap, social circumstances and loneliness in CBS and control patients are given in Table 1. Only with regard to loneliness a statistically significant difference between groups was found. To the table we may add that the percentage of academics was 6% in both groups (3 CBS and 5 control patients). For the 'coping scale', constructed by adding up the scores on the 3 questions on the ability of patients to cope with their visual handicaps, Cronbach's alpha was found to be 0.70. Mean scores on this scale were not significantly different between CBS (7.1, sd 2.6) and control patients (7.3, sd 2.8): $t=0.46$, $p=0.64$).

Table 1. Educational level, social circumstances, loneliness and ability to cope with visual handicap in 50 patients with Charles Bonnet syndrome (CBS) and 80 control patients.

Variable	CBS patients % (N)	Control patients % (N)	χ^2	df	P value
Education					
6 years or less	36 (18)	31 (25)	1.34	2	0.51
7-10 years	30 (15)	40 (32)			
11 years or more	34 (17)	29 (23)			
Social circumstances					
Living alone	54 (27)	37.5 (30)	3.40	1	0.07
Widowed	48 (24)	37.5 (30)	1.40	1	0.24
Social contacts in last 4 weeks:					
0-11	22 (11)	25 (20)	0.78	3	0.86
12-26	32 (16)	25 (20)			
27-42	26 (13)	27.5 (22)			
43 or more	20 (10)	22.5 (18)			
Loneliness					
Score on scale De Jong-Gierveld:					
0-3 = not lonely	36 (18)	65 (52)	11.5	2	0.003
4-6 = moderately lonely	26 (13)	19 (15)			
7-12 = very lonely	38 (19)	16 (13)			
Coping with visual handicap*					
One can live (reasonably) well with eye disease such as mine	58 (29)	71 (57)	2.41	1	0.12
Eyedisease does not (hardly) disturb my joy in life	22 (11)	27.5 (22)	0.49	1	0.48
I have learned to cope (reasonably) well with my eyedisease	76 (38)	72.5 (58)	0.20	1	0.66

* Percentages of patients giving answers indicative of adaptive coping with visual handicap are given in the table. Mean scores on 3-item 'coping scale' are given in the text.

Mean scores on the 5 scales of the NVM personality test are presented in Table 2. In comparison with the NVM norm table for somatic patients (Luteijn & Kok, 1985) mean scores of both CBS and control patients are just below the average range for somatization (11-19) and negativism (12-18), within the average range for shyness (8-15) and within the normal range for psychopathology (0-3). With regard to extraversion mean score of CBS patients is below and of control patients just within the average range (12-17). As shown in the table differences with regard to shyness and extraversion between CBS and control patients were highly significant.

Table 2. Mean scores on NVM-personality test scales of 50 patients with Charles Bonnet syndrome (CBS) and 80 control patients.

NVM scale (score range)	CBS patients		Control patients		T	P value
	mean score	sd	mean score	sd		
Somatization (0-40)	10.2	7.4	9.1	7.4	0.87	0.39
Negativism (0-44)	11.9	6.1	10.3	6.1	1.47	0.14
Extraversion (0-26)	10.0	5.0	12.8	5.4	2.97	0.004
Shyness (0-30)	12.3	6.7	8.6	5.7	3.33	0.001
Psychopathology (0-24)	1.8	2.6	2.1	2.5	-*	0.45

* T test was not applicable because scores on Psychopathology scale had abnormal distribution: large majority of patients scored in range 0-3, indicating "no disturbance". For this reason chi square test was used comparing the percentage of CBS patients (14%) and control patients (23%) with score >3. χ^2 was 1.59 (df=1).

To be able to compare the magnitudes of the predictive effects regarding CBS of loneliness, shyness and low extraversion simultaneously in multiple logistic regression analysis, indicator-variables were created. In accordance with the norm suggested by the authors of the loneliness scale loneliness was defined as a score of >3.²⁰ Following

the NVM norm table 'shyness' was defined as a score of >18 ('high and very high shyness score') and 'low extraversion' as a score of <9 ('low and very low extraversion score').²¹ Thus, 64% of CBS and 35% of control patients were categorized as 'lonely'. Shyness was present in 18% of CBS and 6% of control patients and low extraversion in 46% of CBS and 23% of control patients. The result of the multiple logistic regression analysis is presented in Table 3. In this model 'loneliness' and 'low extraversion' are both significant predictors. 'Loneliness' has the strongest predictive effect. The contribution of 'shyness' is not significant.

Table 3. Results of multiple logistic regression analysis on loneliness, low extraversion and shyness as predictors for the Charles Bonnet syndrome.

Variable	B	SE	P	Exp(B)
Loneliness	1.244	0.394	0.002	3.471
Low Extraversion	0.958	0.434	0.027	2.606
Shyness	0.785	0.664	0.238	2.191
Constant	-1.493	0.330	0.000	

Loneliness: score >3 on loneliness scale De Jong-Gierveld & Kamphuis.²⁰ Low extraversion: score <9 on NVM extraversion scale; shyness: score >18 on NVM shyness scale.²¹

DISCUSSION

As a result of the chosen study design, there were no clinically relevant or statistically significant differences between CBS patients and control patients with regard to age and visual acuity. No support was found for suggestions in literature regarding associations of CBS with a higher level of education, maladaptive coping with visual handicap, or bereavement.

Using the number of social contacts and the living situation of patients as parameters, we found no evidence for connections of CBS with social isolation. However, with regard to loneliness the difference between CBS and control patients was highly significant: the majority of CBS patients felt lonely. Loneliness was common in control patients as well. In view of the advanced age and the severe visual handicap of the study population, the high percentage of lonely people is not surprising.

The NVM personality test showed relatively low mean scores in both groups regarding somatization, negativism and extraversion. This can be explained by the differences in somatic conditions and age between the studied ophthalmic population and the younger population of general somatic patients in which the NVM score ranges

were determined. The authors of the NVM note that scores on negativism and extraversion tend to be lower in elderly patients.²¹ Comparing the personality profiles of both groups the most important findings are the significantly lower extraversion and higher shyness scores in CBS patients. The fact that CBS and control patients did not differ on the psychopathology scale is in line with expectations, as the diagnosis CBS is per definition not compatible with severe psychiatric disorders.

The associations of CBS with loneliness, low extraversion and shyness suggest that CBS is connected with an impairment of social relationships. It seems to be not social isolation proper (as the number of social contacts is not very different in CBS patients and non-hallucinators) but the quality of social contacts that is of importance. Loneliness is the subjective experience of missing significant (high quality) interpersonal relationships. A low score on the NVM extraversion scale indicates that a person assumes a passive attitude towards social contacts. Shyness does not directly imply a low quality of social contacts. This may explain the fact that the association of CBS with shyness is lost if - in multiple logistic regression analysis- loneliness and low extraversion are taken into consideration. Shy people, losing significant contacts as they grow old, may find it difficult to start new personal relationships. However, if they value social contacts they will respond with feelings of loneliness; if not, extraversion is by (NVM) definition low.

Because this was a cross-sectional study we cannot draw definite conclusions on the nature of the relationship between CBS and the associated variables, but a low quality of social contacts could be of causal importance. Several authors noted the disappearance of hallucinations in socially isolated CBS patients after hospitalization. They assume this was due to an increase in the level of sensory stimulation and suggest a lack of stimuli in the home environment contributes to the etiology of CBS.^{7, 13-14, 24} An alternative interpretation of the present study results could be that CBS itself interferes with the social relationships of the patients, as CBS patients may feel embarrassed by their hallucinations. However, this explanation is less likely, because the large majority of patients indicate that CBS causes them little or no distress.⁹

The main conclusion of this study is that loneliness, low extraversion and shyness are risk indicators for CBS in visually handicapped elderly people. Further research will be necessary to examine the nature of the relationship between CBS and the quality of the social contacts of the patients.

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CHAPTER 8

General discussion

INTRODUCTION

In this chapter the study as a whole is critically reviewed. The main conclusions that can be drawn from this study are discussed. CBS is compared with hallucinations in psychotic disorders. And finally, we will go into the directions that future research on the CBS could take.

CRITICAL REVIEW

Diagnostic criteria for CBS

There are no generally accepted diagnostic criteria for CBS. The authors of the various definitions agree on the inclusion criterion “complex visual hallucinations” and the fact that hallucinations in the context of (another) psychiatric disorder should not be included in the diagnosis. However, there has been much discussion on the question whether or not several somatic disorders should be considered as additional exclusion criteria. In view of this discussion and the lack of research on connections between isolated visual hallucinations and somatic disorders, Gold and Rabins proposed diagnostic criteria based on clinical characteristics, discriminating CBS from other psychiatric syndromes, but leaving questions about etiology (e.g. eye pathology or brain lesions) to future study.¹ The criteria used in the present study are based on the criteria of Gold and Rabins. There are two differences: first, the original criterion “hallucinations are stereotyped” was omitted because we found insufficient support for this characteristic in literature (see chapters 2 and 3); second, we operationalized the criterion “hallucinations are persistent or repetitive” introducing a minimum duration of the syndrome of 4 weeks. Thus, the following criteria were used to include patients: a) at least one complex visual hallucination within the 4 weeks prior to screening; b) a period between the first and the last hallucination exceeding 4 weeks; c) full or partial retention of insight into the unreal nature of the hallucinations; d) absence of hallucinations in other sensory modalities; e) absence of delusions.

In the populations we studied, the above diagnostic criteria served their purpose: in CBS patients, complex visual hallucinations were indeed “isolated” psychopathological phenomena. Nevertheless, it became clear to us that the criteria are far from perfect.

The criterion ‘hallucinations are persistent or repetitive’ (which we operationalized by using a minimum duration of the syndrome of 4 weeks) was proposed by Gold & Rabins to “distinguish CBS from phenomena such as the hallucinations of widowhood”.¹ It is however somewhat unsatisfactory to exclude patients, with in all other respects typical characteristics of CBS, for the sole reason that their hallucinatory experiences did not last long enough. Furthermore, it is doubtful whether this criterion is helpful: hallucinations of widowhood may also occur repeatedly over a longer period of time.

The criterion “full or partial insight” is ment to exclude psychotic patients. This

raises the question how “partial” insight may be. Insight some time after the hallucinatory episode does not suffice, because many patients may just as well become aware of the unreal nature of their hallucinations after a true psychotic episode. As mentioned in chapter 6 we would favor changing this criterion to “full insight”.

Another criterion with the purpose to exclude psychosis is “absence of hallucinations in other sensory modalities”. There are however patients who experience complex visual hallucinations as well as “musical hallucinations” (a syndrome often considered to be the acoustic counterpart of CBS) with retention of insight.²⁻³ These patients are wrongfully excluded, because they are clearly not suffering from psychosis.

One could also argue that the diagnostic criteria, aiming only to exclude other psychiatric disorders, are too broad. In a recent review article Fernandez and co-workers list the following proposed exclusion criteria: dementia, delirium, focal cerebral lesions, Parkinson’s disease and levo-dopa induced hallucinations, organic mood syndromes, intoxications, lesions of the central visual pathways or cortex and acute eye disease.⁴ We do not agree with these exclusion criteria. It is not necessary to explicitly mention dementia and delirium as exclusion criteria, because it is unlikely that delirious or demented patients meet the present diagnostic criteria for CBS (insight into the unreal nature of visual hallucinations; absence of delusions).⁵ With regard to the other disorders on the list of Fernandez et al, it should be remembered that only small minorities of patients with these conditions have visual hallucinations. For that reason, the presence of these disorders does not explain the occurrence of hallucinations. Like the visual handicap of the present study population they may merely be factors contributing to the occurrence of hallucinations. Furthermore, whether or not hallucinations in patients with these disorders meet CBS criteria has not been subject of systematic research. However, there are situations, in which complex visual hallucinations (with insight) may occur as integrated and understood symptoms of other conditions. These conditions should indeed exclude patients from the diagnosis CBS. This applies to: a) ‘hallucinations of widowhood’ (as mentioned above the present ‘time-criterion’ does not suffice as an exclusion criterion); b) complex visual hallucinations which, as symptoms of epileptic or migrainous attacks, are considered to be ‘irritative phenomena’; c) hypnagogic and hypnopompic hallucinations; d) hallucinations occurring in exceptional situations (extreme forms of stress and/or social isolation, such as in hostages); e) hallucinations due to hallucinogenic drugs (LSD, mescaline).

Summarizing the above considerations, we think the following diagnostic criteria would be an improvement: a) presence of complex visual hallucinations; b) intact reality testing in all respects (including full insight into the unreal nature of hallucinations and absence of delusions); c) exclusion of hallucinations of widowhood, hypnagogic and hypnopompic hallucinations, hallucinations secondary to epileptic or migrainous attacks, hallucinations due to hallucinogenic drugs and hallucinations occurring in exceptional stressful situations.

Clinical characteristics

Clinical characteristics were topics in chapters 3 and 5. Not all interesting characteristics were studied extensively. For instance, some circumstances favoring the occurrence of hallucinations and techniques to suppress hallucinations were mentioned by several patients spontaneously. The prevalence of these characteristics remains unclear because not all patients were systematically interviewed on them. The course of CBS was studied retrospectively with all hazards associated with such an approach.

Prevalence and risk indicators

The prevalence of CBS and risk indicators for CBS in visually handicapped patients were the main issues in chapters 4, 6 and 7. The prevalence of CBS in this population was high enough to pursue the aims of the study. An important reason for the high participation rate is probably that most of the study took place at home with the patients. We are convinced that the high prevalence of CBS we found is not an overestimation, as the diagnosis was not based on the screening interview only, but was thoroughly checked in a full psychiatric examination. Possibly, the prevalence found is an underestimation. As the semistructured interview we used is not validated, some CBS patients may not have understood the questions asked or may have been afraid to admit they had hallucinations.

Associations of CBS with advanced age and low visual acuity were clearly demonstrated in comparative prevalence studies in young and old low vision patients and elderly patients with different levels of visual acuity.

Studying the medical, psychological and social profile of CBS patients we used the term "risk indicators" for factors associated with CBS. We avoided the term "risk factor", because the cross sectional design of this part of the study prevents us from drawing the conclusion that these associated factors increase the risk of developing CBS. The presence of a risk indicator merely makes it more likely that CBS is present.

The population, in which possible risk indicators were examined, is not representative of the general population of visually handicapped people: in order to be referred to the low vision unit people have to be motivated to make the best of their handicap and be physically and mentally fit to visit this out patients unit. For that reason, the findings should not, without due consideration, be applied to other populations.

Because little systematic research on CBS had been done in the past, we had no convincing indications which possible risk indicators were the most promising. For that reason, we chose to study a broad range of variables. A limitation of this part of the study is that several screening methods were rather superficial: the Mini Mental State Examination, which was used to determine cognitive functions, is known to lack sensitivity and has disadvantages if applied to visually handicapped people (because some tasks are disturbed by bad eye sight).⁶ It would have been better to use a more sensitive instrument, more so because severe cognitive disorders were rare in this population. One could have doubts regarding the interview on the ability of patients to

cope with their visual handicap. Lacking a useful Dutch language test we developed a provisory scale ourselves, which is not yet validated. The impact of the findings is limited as well by the fact that no objective methods were used to examine biological factors, such as neuro-imaging techniques and blood tests. Finally, the cross-sectional design of the study, does not allow us to draw definite conclusions on possible causal relationships between risk-indicators and CBS.

MAJOR CONCLUSIONS

Finding a high prevalence of CBS in visually handicapped people, the long held assumption that CBS is rare was proven wrong. Admittedly, there have been two earlier, smaller, studies indicative of such a prevalence in comparable populations.⁷⁻⁸ In the present study statistical support for connections of CBS with low visual acuity and advanced age was found.

The clinical features of CBS were systematically studied in a large number of patients. They proved to be variable, though some characteristics were shared by the majority of patients. Several findings, including those on factors triggering and stopping hallucinations, may be clues to the pathogenesis of CBS. Other findings raise intriguing questions. For instance, what is the implication of the fact that some patients indicate they see 'images from the past' as well as images they do not remember ever having seen in reality? Are there two kinds of hallucinations in CBS: one kind resulting from the reappearance of earlier normal perceptions and another involving "new" images resulting from an unconscious creative process? Or have the patients forgotten they once encountered the hallucinated objects in reality?

The finding of risk indicators -loss of energy, compromised general health, β -blocking medication, loneliness, low extraversion and shyness- in elderly low vision patients provides a starting point for future research on possible causal factors. The present theories on the etiology of CBS (reviewed in chapter 2) are based on little empirical data and are not convincing. For the psychological theory which placed wishfulfillment in the centre of attention no support was found in this study. Theories, emphasizing eye disease or visual handicap as the cause, fail to explain the fact that only a minority of low vision patients have CBS and that CBS has been found in patients with intact vision as well. Theories which imply a specific cerebral disorder leave questions on the connection between CBS and low vision unanswered. Probably, there are many factors which contribute to the occurrence of CBS. Further research will be necessary to gather the building stones for a theory on a multifactorial etiology of CBS.

CBS AND PSYCHOSIS

The CBS is not a major mental health problem, but it provides an excellent opportunity to study hallucinations, because in this syndrome they are -from a psychiatric point of view- isolated phenomena.

However, it remains to be seen to what extent the hallucinations of CBS patients and those of truly psychotic patients are similar phenomena. The extraordinary perceptions described by the CBS patients in this study meet the DSM-IV definition of hallucinations (see chapter 2).¹¹ Still, some critics argued the term “pseudohallucinations” should have been used, in view of the retention of insight. However, the term “pseudohallucinations” is ambiguous: some authors describe pseudohallucinations as forms of imagery experienced in inner subjective space. Others define pseudohallucinations as a particular variety of hallucinations: those accompanied by insight (either current or retrospectively). Furthermore, if the latter definition is used, one is confronted with the problem that insight is not an all-or-none category: it may be fluctuating and partial. For those reasons, it is recommended in literature to refrain from the use of the term “pseudohallucinations”. The term “hallucinations” should be applied to phenomena such as in CBS and the degree of insight mentioned.⁹⁻¹⁰ We followed this recommendation. Nevertheless, the presence of insight discriminates CBS from psychosis. Perhaps, the presence or absence of insight should not be viewed as a feature of the hallucinatory experiences themselves, but as a reflection of the state of the patients’ executive functions. In CBS executive functions are intact: though the perceptions are abnormal, the interpretation is normal. In psychosis a more global cerebral disturbance interferes with reality testing, which leads to an abnormal interpretation of normal perceptions (resulting in delusions) as well as abnormal perceptions (hallucinations with loss of insight).

Apart from the issue of insight, there are several other (relative) differences. The large majority of CBS patients experience non-threatening hallucinations, whereas psychotic patients often feel terrified by their experiences. Partly, this could be connected with differences regarding reality testing. However, the content of hallucinations often seems to be more harmless in CBS compared to psychosis. Furthermore, most CBS patients indicate their hallucinations lack a personal relevance. It is our clinical impression -we found no systematic studies on the subject- that the content of hallucinations in delirium and dementia often lack a personal relevance as well, whereas schizophrenic patients tend to experience most relevant hallucinations (e.g. voices talking to or about the patient).

Summarizing, we have to conclude that the relationship between patients and their hallucinations is different in CBS and psychosis. It is not yet clear whether this is due to a different nature of the hallucinations themselves or that this results from additional cerebral disturbances in psychotic patients.

RECOMMENDATIONS REGARDING FUTURE RESEARCH

The present study leaves many questions unanswered and raises new questions. Some authors consider CBS to be an early marker for dementia.¹² We found no associations between CBS and cognitive deficits in a cross-sectional case-control study. However, a long term follow up study is necessary to determine whether CBS increases the risk for dementia. Hypotheses on possible etiological roles of risk indicators can be tested. The effectiveness of hallucination-suppressing acts can be studied systematically.

There are several other promising topics for future research. Modern neuro-imaging techniques could help to localize the cerebral centres involved. The therapeutic values of carbamazepine, melperone and andazolone, medications which seemed to be effective in case studies, can be tested.¹³⁻¹⁴

Though the studies proposed above may help to gain more insight into the CBS, the road to a real understanding of the phenomenon may still be long. In 1760 Charles Bonnet wrote that it would take at least two centuries before scientists could reveal all its secrets.¹⁵ In his time Bonnet was considered to be a pessimist.¹⁶ However, we may now conclude he was -at least in one respect- a realist: in 1998 CBS still is largely a mystery.

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SUMMARY

The Charles Bonnet syndrome (CBS) is characterized by the presence of complex visual hallucinations in psychologically normal people. Although this phenomenon was recognized more than 200 years ago, little systematic scientific research has been done on the subject. Possible reasons for this are the alledged rarity of the syndrome and the lack of generally accepted diagnostic criteria. The present knowledge on the syndrome is based almost entirely on case studies. This thesis comprises an explorative systematic empirical study of the CBS.

Chapter 1 starts with an introduction on hallucinations in general and the CBS. The aim and outline of this study are delineated.

In *chapter 2* a typical case is described and the literature on CBS since 1760 is reviewed. It proves that, apart from the fact that most patients demonstrate insight into the unreal nature of their hallucinations, the psychopathological characteristics of the CBS vary from case to case. Based on casuistic data the following risk factors are suggested: advanced age, female sex, disorders of the visual system, maladaptive coping with visual loss, bereavement, and social isolation. Indications are found that CBS could be relatively common in elderly ophthalmic patients. Various theories on etiology and pathogenesis are discussed. It is noted that these theories lack a sound empirical basis. With regard to the definition of CBS, the following diagnostic criteria (largely based on a proposal of Gold & Rabins in 1989) are suggested: 1) presence of formed and complex, persistent or repetitive visual hallucinations; 2) full or partial retention of insight into the unreal nature of these hallucinations; 3) absence of primary or secondary delusions; 4) absence of hallucinations in other sensory modalities.

Chapter 3 comprises a report on a comprehensive study on the characteristics of the CBS and psychiatric, neurologic and ophthalmologic findings in a series of 14 patients. It proves feasible to discriminate CBS from other (psychiatric) syndromes by the diagnostic criteria that were suggested in chapter 2. Common characteristics of hallucinations include the absence of a personal meaning of the content of hallucinations and the disappearance of hallucinations after eye closure. There is no evidence for a relationship of the syndrome with psychiatric disorders. In the majority of cases, ocular pathology and neurologic disturbances are found. Most patients in this series also suffer from social isolation.

Based on these data a possible multifactorial etiology of CBS is discussed.

In *chapter 4* the results of a prevalence study on CBS in low vision patients and elderly general ophthalmic patients are given. All subjects are patients from the Department of Ophthalmology of the University Hospital Nijmegen. A semi-structured interview on visual hallucinations is administered to 300 adult patients from the low-vision unit and 200 general ophthalmic patients who had been referred for optic refraction measurements. Positive cases are screened with the Geriatric Mental State Schedule and Mini Mental State Examination. Ophthalmic and sociodemographic data are

gathered for all patients. The prevalence of CBS in patients from the low-vision unit is 11%. A prevalence of only 1% is found in the elderly general ophthalmic patients. CBS is significantly associated with an age over 64 years and a visual acuity in the better eye of 0.3 or less. No significant associations with ophthalmic diagnoses, patient sex, marital status, or social circumstances are found.

It is concluded that these findings support connections of CBS with advanced age and sensory deprivation.

In *chapter 5* an extensive description of the clinical characteristics of CBS is given. Sixty of 505 patients from the low vision unit of the Department of Ophthalmology of the University Hospital Nijmegen, who participated in a case-finding study, meet criteria for CBS. All patients are interviewed on the psychopathological characteristics, the personal meaning and the emotional impact of hallucinations, as well as on factors influencing hallucinations. It is found that, although diagnostic criteria demand merely 'partial insight', all patients have full insight into the unreal nature of their hallucinations. Other characteristics vary. In 46 (77%) patients, hallucinations always lacked a personal meaning. Sensory deprivation and a low level of arousal seem to favour the occurrence of hallucinations. CBS causes considerable distress in only 17 (28%) patients. However, most patients are relieved to be informed about the fact that their hallucinations are not due to mental disease. In only one of the 16 patients who had consulted a doctor had the proper diagnosis been made.

It is concluded from these findings that CBS should be considered as a diagnosis in patients who complain of visual hallucinations. Though there is no proven treatment, many patients will benefit from reassurance that their hallucinations do not imply mental illness.

In *chapter 6* the results of a case-control study on possible associations of CBS with somatic and psychiatric factors are given. All subjects are patients over the age of 64 years from the low vision unit of the Department of Ophthalmology of the University Hospital Nijmegen. Fifty-two patients with CBS and eighty patients without visual hallucinations are interviewed, using a checklist on medical history, current medical complaints, medication, and hearing loss; Mini Mental State Examination; and Geriatric Mental State Schedule. Significant associations of CBS with the number of current somatic disorders, the use of β -blocking medication, and (subjective) loss of energy are found. There are no significant differences between CBS patients and controls with regard to cerebral and metabolic disorders, hearing loss, cognitive status, and psychiatric disorders.

These findings suggest that the use of β -blocking medication and subjective loss of energy -probably connected with compromised general health- are risk factors for CBS.

Chapter 7 comprises a case-control study on connections of CBS with social and psychological factors. All subjects are patients over the age of 64 years from the low

vision unit of the Department of Ophthalmology of the University Hospital Nijmegen. Fifty patients with CBS and eighty patients without visual hallucinations are interviewed on their educational level, social circumstances, number of social contacts and ability to cope with their visual handicap. Loneliness is measured with the loneliness scale of De Jong-Gierveld and personality traits are examined with the NVM personality test. Compared to the control group, significantly more CBS patients are lonely. Mean scores on NVM shyness scale are significantly higher and on NVM extraversion scale significantly lower in CBS patients. Using multiple logistic regression analysis with these 3 determinants, dichotomised as indicator variables, simultaneously, 'loneliness' and 'low extraversion' prove to be significant predictors of comparable magnitudes. In this model the effect of 'shyness' is not significant.

It is concluded that loneliness, low extraversion and shyness are risk indicators for CBS in elderly visually handicapped people. These findings suggest a connection between CBS and a low quality of social contacts.

Chapter 8 comprises a general discussion on the study as a whole. The methodological aspects of the study are critically reviewed. Shortcomings of the present diagnostic criteria are noted and a proposal to improve these criteria is given. The limitations of the study, due to the specific population characteristics, the cross sectional design of the study on risk indicators, and the screening instruments used, are discussed.

The most important results of the study are mentioned: a high prevalence of the CBS in elderly visually handicapped patients was found; the clinical characteristics of CBS were delineated in a large group of patients; psychological, social and biological risk indicators for CBS were detected.

In a comparison of CBS with psychosis, the differences and similarities of hallucinations in CBS, schizophrenia, dementia and delirium are discussed.

Finally, recommendations regarding future research on CBS are given.

SAMENVATTING

Het syndroom van Charles Bonnet (CBS) wordt gekenmerkt door het optreden van complexe visuele hallucinaties bij geestelijk gezonde personen. Hoewel dit verschijnsel reeds meer dan 200 jaar bekend is, is er weinig systematisch wetenschappelijk onderzoek naar verricht. Mogelijke redenen daarvoor zijn de vooronderstelde zeldzaamheid van het syndroom en het ontbreken van algemeen geaccepteerde diagnostische criteria. De huidige kennis over het syndroom berust bijna geheel op casuïstische mededelingen. Dit proefschrift behelst een verkennend, systematisch empirisch onderzoek naar het CBS.

In *hoofdstuk 1* wordt een inleiding gegeven betreffende het verschijnsel hallucinatie in het algemeen en het CBS in het bijzonder. Doel en opzet van deze studie worden uiteengezet.

In *hoofdstuk 2* wordt een kenmerkende casus beschreven en een overzicht gegeven van de wetenschappelijke literatuur betreffende het syndroom sinds 1760. Het blijkt dat, afgezien van het feit dat de meeste patiënten inzicht toonden in het irële karakter van hun hallucinaties, de psychopathologische kenmerken van het CBS varieerden van casus tot casus. Op grond van casuïstische bevindingen worden de volgende risico factoren gesuggereerd: hoge leeftijd, het vrouwelijk geslacht, aandoeningen van het visuele systeem, gestoorde coping met visusverlies, verlieservaringen en sociaal isolement. Er zijn aanwijzingen dat het CBS in de mogelijke risico groep van oudere oogheelkundige patiënten minder zeldzaam is dan lange tijd werd aangenomen. Diverse theorieën over aetiologie en pathogenese worden besproken, met de aantekening dat deze empirisch onvoldoende onderbouwd zijn. Met betrekking tot de definitie van het CBS worden de volgende diagnostische criteria (grotendeels gebaseerd op een voorstel van Gold & Rabins in 1989) aangegeven: 1) de aanwezigheid van gevormde, complexe, persisterende of herhaald optredende visuele hallucinaties; 2) geheel of gedeeltelijk behoud van inzicht in het irële karakter van de hallucinaties; 3) afwezigheid van primaire of secundaire wanen; 4) afwezigheid van hallucinaties in andere zintuiglijke modaliteiten.

In *hoofdstuk 3* wordt verslag gedaan van een uitgebreid onderzoek naar de kenmerken van het CBS en psychiatrische, neurologische en ophthalmologische bevindingen bij 14 patiënten met het syndroom van Charles Bonnet. Het blijkt mogelijk het CBS met behulp van de in hoofdstuk 2 genoemde diagnostische criteria te onderscheiden van andere (psychiatrische) syndromen. Bij alle patiënten wordt vastgesteld dat de inhoud van de hallucinaties een persoonlijke relevantie ontbeert en dat de hallucinaties verdwijnen als de ogen worden gesloten. Er zijn geen aanwijzingen voor een relatie van het CBS met psychiatrische stoornissen. De meeste patiënten hebben oogheelkundige en neurologische aandoeningen. Tevens verkeerden de meesten in een sociaal isolement.

De conclusie van dit onderzoek is dat er waarschijnlijk een combinatie van aetiologische factoren aan het CBS ten grondslag ligt.

Hoofdstuk 4 behelst een prevalentie onderzoek naar het CBS bij low-vision patiënten en oudere oogheelkundige patiënten in het algemeen. Alle deelnemers aan dit onderzoek zijn patiënten van de afdeling Oogheelkunde van het Academisch Ziekenhuis Nijmegen. Driehonderd volwassen patiënten van de low vision polikliniek en 200 patiënten, ouder dan 64 jaar, die ter bepaling van de refractie waren verwezen naar de optometrie polikliniek ondergaan een semi-gestruktuureerd interview betreffende visuele hallucinaties. Patiënten met hallucinaties werden onderzocht met de Geriatric Mental State Schedule en de Mini Mental State Examination. Oogheelkundige en sociodemografische gegevens worden verzameld bij alle patiënten. In de populatie van de low-vision polikliniek is de prevalentie van het CBS 11% . De prevalentie bij de patiënten van de optometrie polikliniek is slechts 1%. Statistisch significante associaties van het CBS met een leeftijd boven de 64 jaar en een visus van het beste oog van 0,3 of lager worden vastgesteld. Er zijn geen significante associaties met oogheelkundige diagnose, geslacht, burgerlijke staat, of het al dan niet alleen wonen van de patiënten.

Geconcludeerd wordt dat de bevindingen steun geven aan de vooronderstellingen dat het CBS samenhangt met sensorische deprivatie en een hoge leeftijd.

Hoofdstuk 5 is gewijd aan een uitgebreide beschrijving van de klinische kenmerken van het CBS. Van de 505 patiënten die deelnamen aan een screenings onderzoek op de low vision polikliniek van de polikliniek oogheelkunde voldeden er 60 aan de criteria voor het CBS. Deze patiënten worden geïnterviewd met betrekking tot de psychopathologische kenmerken, de persoonlijke betekenis en de emotionele impact van de hallucinaties, alsmede factoren die van invloed zijn op de hallucinaties. Er wordt vastgesteld dat, hoewel in de diagnostische criteria “partieel inzicht in het irrealistische karakter van de hallucinaties” voldoende werd geacht, bij alle patiënten sprake is van volledig inzicht. De overige psychopathologische kenmerken zijn variabel. Bij 46 (78%) patiënten ontbeerden de hallucinaties altijd een persoonlijke relevantie. Sensorische deprivatie en een laag niveau van waakzaamheid lijken het optreden van hallucinaties te bevorderen. Noemenswaardige lijdensdruk veroorzaakt het CBS slechts bij 28% van de patiënten. Desondanks zijn de meeste patiënten opgelucht te horen dat hun hallucinaties niet veroorzaakt worden door een geestesziekte. De juiste diagnose was voorheen slechts gesteld bij één van de 16 patiënten die een arts hadden geraadpleegd.

Uit deze bevindingen volgt de aanbeveling dat de diagnose CBS in overweging dient te worden genomen bij patiënten die klagen over visuele hallucinaties. Hoewel er nog geen behandeling met bewezen effectiviteit bestaat kunnen vele patiënten profiteren van de geruststelling dat hun hallucinaties geen geestesziekte impliceren.

In *hoofdstuk 6* worden de resultaten gegeven van een case-controle onderzoek naar de samenhang van het CBS met somatische en psychiatrische factoren. Alle patiënten zijn ouder dan 64 jaar en afkomstig van de low-vision polikliniek van de afdeling Oogheelkunde van het Academisch Ziekenhuis Nijmegen. Twee-en-vijftig patiënten

met CBS en 80 patiënten zonder visuele hallucinaties worden geïnterviewd, gebruikmakend van een checklist met betrekking tot de medische voorgeschiedenis, actuele medische klachten, medicatie en gehoorsproblemen; de “Mini Mental State Examination” en de “Geriatric Mental State Schedule”. Een statistisch significante samenhang van het CBS met het aantal actuele somatische ziekten, het gebruik van β -blokkerende medicijnen en een (subjectief) tekort aan energie wordt vastgesteld. Er zijn geen significante verschillen tussen patiënten met het CBS en controle patiënten met betrekking tot cerebrale en metabole stoornissen, gehoorsvermindering, cognitie en psychiatrische stoornissen.

Deze bevindingen suggereren dat het gebruik van β -blokkerende medicijnen en een subjectief tekort aan energie -mogelijk verbonden met een verzwakte algemene gezondheid- risicofactoren zijn voor het CBS.

In *hoofdstuk 7* wordt verslag gedaan van een case-controle onderzoek gericht op de samenhang van het CBS met sociale en psychologische factoren. Alle patiënten zijn ouder dan 64 jaar en afkomstig van de low-vision polikliniek van de afdeling Oogheelkunde van het Academisch Ziekenhuis Nijmegen. Vijftig patiënten met het CBS en 80 patiënten zonder visuele hallucinaties worden geïnterviewd met betrekking tot hun opleidingsniveau, woonomstandigheden, aantal sociale contacten en hun vermogen om te gaan met hun visuele handicap. Eenzaamheid wordt gemeten met de eenzaamheidsschaal van De Jong-Gierveld & Kamphuis en de persoonlijkheid wordt getest met de Nederlandse Verkorte MMPI (NVM). Vergeleken met de controle groep zijn significant meer patiënten met het CBS eenzaam. De gemiddelde scores op de NVM extraversie schaal zijn significant lager en op de NVM verlegenheidsschaal significant hoger in de CBS groep. In de multiële logistische regressie analyse van deze 3 determinanten, in de vorm van indicator variabelen, blijken “eenzaamheid” en “lage extraversie score” significante predictoren met een vergelijkbaar risico-verhogend effect. In dit model is het effect van “verlegenheid” niet significant.

De conclusie is dat eenzaamheid, verlegenheid en een laag niveau van extraversie risico-indicatoren zijn voor het CBS bij visueel gehandicapte ouderen.

Hoofdstuk 8 behelst een algemene discussie met betrekking tot de studie als geheel.

De methodologische aspecten van de studie worden kritisch beschouwd. De tekortkomingen van de gebruikte diagnostische criteria voor het CBS worden belicht en een voorstel voor verbetering wordt gedaan. De beperkingen van de studie, die voortvloeien uit de specifieke populatie-kenmerken, het cross sectionele design van het onderzoek naar risico-indicatoren en de gebruikte onderzoeksinstrumenten, worden besproken.

Als belangrijke resultaten van de studie worden genoemd: de hoge prevalentie van CBS bij oudere visueel gehandicapte mensen, het in kaart brengen van de klinische kenmerken van het syndroom bij een grote groep patiënten en de vaststelling van psychische, sociale en biologische risico-indicatoren voor het CBS.

Het CBS wordt vergeleken met psychosen, waarbij overeenkomsten en verschillen

tussen het CBS en hallucinaties in het kader van schizofrenie, dementie en delirium worden besproken.

Ten slotte worden enige suggesties voor verder wetenschappelijk onderzoek gegeven.

APPENDIX

STUDY INSTRUMENTS

In this study several wellknown instruments were used: Geriatric Mental State Schedule, Mini Mental State Examination, Loneliness scale De Jong-Gierveld and Nederlandse Verkorte MMPI (NVM). For information on these instruments we refer to the publications mentioned in the lists of references. The following instruments were especially developed for the purpose of this study and are presented in this appendix: I. screening form spontaneous visual phenomena; II. questionnaire clinical characteristics Charles Bonnet syndrome; III. checklist somatic disorders and medication; and IV. questionnaire on coping with visual handicap.

I. SCREENING FORM SPONTANEOUS VISUAL PHENOMENA

PATIENT NUMBER:

DATE SCREENING:

SCREENER:

A. DEMOGRAPHIC DATA

name:	initials:
birth date:	sex: m/f
address:	
phone:	
ophthalmologist:	
GP:	phone:
marital status: married/wid./not marr./div. (since)	
living situation: independent/home for elderly/nursery home/ other:	
living alone: yes / no	
B. OPHTHALMIC DATA	
diagnoses OD:	
diagnoses OS:	
Visual acuity	OD: date: OS:

Informed consent

Dear Madam / Sir, people with impaired vision sometimes perceive things differently or see thing they did not see before. It is not yet known how often this is the case. In the context of a scientific investigation I would like to ask you some questions about this. It will take about 10 minutes of your time. Your participation is voluntarily. If you don't want to participate, your treatment will not be influenced in a negative way. May I ask you the questions?

Yes/No

C. QUESTIONNAIRE SPONTANEOUS VISUAL PHENOMENA

Score: no=0; yes=1; unclear=8

1. Do you see things or persons different from how they really are, for instance larger, smaller, further away, or closer?	0	1	8
2. Are the shapes of things or persons distorted?	0	1	8
3. Do you see things or persons with extraordinary sharpness or brightness?	0	1	8
4. Do things look peculiar in some other way?	0	1	8
5. Do you see spots, starlets, stripes, flashes of light, or similar things?	0	1	8
6. Have you at some time been deceived by your imagination?	0	1	8
7. Do you sometimes mistake one thing for another?	0	1	8
8. Do you see strange things?	0	1	8
Comment on questions 1-8: description, occurrence in last 4 weeks:			
9. Have you seen things other people could not see?	0	1	8
10. Have you seen things which were not really present?	0	1	8
11. Have you experienced visions?	0	1	8
(Question to relative/aquaintance accompanying patient, if appropriate)			
12. Have you noticed (name patient) saw things which were not perceived by others?	0	1	8
(Observation screener)			
13. Patient mentions possible complex visual hallucinations outside of the context of questions 9-12.	0	1	8
If score 0 on all items 9-13: no case = end questionnaire. If score 1 or 8 on one of the items 9-13: possible case = continue with question 14.			
Judgement screener: possible case?	0	1	8

14. Can you describe the things you have seen?			
Judgement screener: are perceptions formed and complex?	0	1	8
15. Is it possible you misinterpreted things that were really present? (for instance mistook a vase for a face) Judgement screener: perceptions are not illusions?	0	1	8
16. Do you see these things in the surrounding space? (in the environment, not only in the mind, not in the imagination)	0	1	8
17. Can you evoke these perceptions by your own free will? Judgment screener: are perceptions independent of the patients will?	0	1	8
18. Are the things you see really the things you say they are? (or are they just resembling those things, are they vague images or spots)	0	1	8
19. Are you awake when you see these things?	0	1	8
20. Have you seen these things in the last 4 weeks?	0	1	8
Questions to relative/aquaintance accompanying patients (if appropriate)			
21. Have you been present at some time that (name patient) saw something you did not see?	0	1	8
22. Do you think he/she misinterpreted something/ someone really present?	0	1	8
23. Did he/she think that something/someone on television was present in the room?	0	1	8
24. Did he/she think his/her mirror image was another person?	0	1	8
If score 1 on all items 14-20 and score 0 on all items 22-24: patient is a case (=complex visual hallucinations in last 4 weeks). If score 0 on one of items 14-20, and/or score 1 on one of the items 22-24: no case.			
25. Judgement screener: is patient a case?	0	1	8

II. CLINICAL CHARACTERISTICS CHARLES BONNET SYNDROME

1. Duration of syndrome:	
2. Frequency of hallucinations:	
3. Duration of hallucinations:	
4. Duration periodes with hallucinations: Duration periodes without hallucinations:	
5. Formal aspects spontaneous visual phenomena	
a) localization in visual field:	
b) fitting well in surroundings?	Yes/no/sometimes
c) moving along with eyes?	Yes/ no/sometimes
d) perceived with which eye?	Left/right/both
e) intrinsic movements?	Yes/no/sometimes
f) movement 'en bloc'?	Yes/no/?
g) always same images?	Yes/no/?
h) magnitude and position images always the same?	Yes/no/?
i) colouring?	Coloured/black-white
j) clearness compared to normal perceptions?	Clearer, less clear, equalr
6. Recognition objects from former normal perceptions?	Yes/no/sometimes
7. Circumstances favouring occurrence of hallucinations:	
a) morning/afternoon/evening/night	
b) daylight/artificial light/dusk-dawn/darkness	
c) at rest/ engaged in activities/ both	
d) at home/ elsewhere/ both	
e) alone/ in company/ both	f) other circumstances: ...
8. Affective response to hallucinations	
a) dominating affect: ...	
b) anxiety	sadness
irritability	amazement
amused	joy
indifference	
9. Voluntary influence on hallucinations	
a) Patient can voluntarily evoke hallucinations?	Yes/no
How? ...	
b) Patient can voluntarily stop hallucinations by ..	
-closing/opening the eyes	Yes/no
-approaching hallucinated object	Yes/no
-otherwise: ...	Yes/no
c) Patient can influence content of hallucinations?	Yes/no

- | | |
|---|--------|
| 10. Insight into unreal nature while hallucinating? | Yes/no |
| 11. Connection with headache or epilepsy? | Yes/no |
| 12. Connection with waking up/falling asleep? | Yes/no |
| 13. Well awake when hallucinating? | Yes/no |
| 14. a) Are hallucinations a burden to the patient? | Yes/no |
| b) Would patient take medication to suppress hallucinations? | Yes/no |
| 15. Has patient consulted a doctor because of hallucinations?
Why / why not: ... | Yes/no |

III . CHECKLIST SOMATIC DISORDERS AND MEDICATION

Anamnesis

1. Disorders/ diseases/ operations	Date

checklist:

heartdisease	high blood pressure	diabetes
stroke	cerebral haemorrhage	Parkinson
thyreoid disease	epilepsy	headaches/migraine
renal disease	liver disease	brain injuries

2. Have you ever suffered from a sudden paralysis of a limb or a speech disorder?
If yes, when did this happen? ...

3. Actual fysical complaints yes/no
Specification: ...

4. Medications

name	Dose	Frequency	starting date

5. In case of neurological disorder:
diagnosis, symptoms and course

FYSICAL EXAMINATION

General impression: ...

Pulse: ... reg/irreg

RR: ... / ... mm Hg

Hearing problems: ...

Auscultation

Heart: ...

Carotic arteries: ...

Lungs: ...

Conjugated eye movements: ...

Facial expression: ...

Gait: ...

Tremor: ...

Diadochokinesis: ...

Muscular power

-Facial:

-Arms:

-Legs:

Muscle tone

Velocity

Reflexes

-snout: ...

-jaw: ...

palmomental: ...

-biceps: ...

-supinator: ...

-knee: ...

-ankle: ...

-plantar: ...

Further notes:

IV. COPING WITH EYE DISEASE

- | | | |
|---|---|--------------|
| 1. Can a person live with an eye disease such as yours? | 0 | not |
| | 0 | hardly |
| | 0 | more or less |
| | 0 | reasonably |
| | 0 | well |
| 2. To what extent does your eye disease disturb your joy in life? | 0 | not |
| | 0 | somewhat |
| | 0 | more or less |
| | 0 | rather |
| | 0 | severely |
| 3. Have you learned to cope with your eye disease? | 0 | no |
| | 0 | hardly |
| | 0 | more or less |
| | 0 | reasonably |
| | 0 | yes |

Dankwoord

Het wetenschappelijk onderzoek dat ten grondslag ligt aan dit proefschrift is ontsproten uit de klinische praktijk: in 1991 meldde zich een oudere dame op mijn spreekuur omdat zij sinds enige jaren miniatuur-schoorsteenvegers zag rondlopen in haar huiskamer. Zij was zich terdege bewust van het irrecële karakter van haar uitzonderlijke waarnemingen. Psychiatrisch onderzoek bracht geen gegevens aan het licht die het mij mogelijk maakten de verschijnselen fatsoenlijk in een mij bekende diagnostische categorie onder te brengen. Uiteindelijk leidden naspeuringen in de literatuur toch tot de diagnose: het zeldzame syndroom van Charles Bonnet. Toen ik een paar maanden later opnieuw met dit syndroom geconfronteerd werd, raakte ik geïntrigeerd, begon te twijfelen aan de “zeldzaamheid” en vatte het plan op een serie patiënten te gaan verzamelen. Het lukte een voldoende aantal patiënten te vinden, te onderzoeken en de bevindingen te publiceren. Dat motiveerde tot het opzetten van een groter wetenschappelijk onderzoek, hetgeen ten slotte uitmondde in dit proefschrift. Daarvoor, dat het zover is gekomen, ben ik aan vele mensen dank verschuldigd.

Bij het wetenschappelijk onderzoek ben ik begeleid door een aantal deskundigen. Mijn erkentelijkheid gaat uit naar Prof. Dr. F.G. Zitman, voor zijn niet-aflatende inzet als mijn voornaamste wetenschappelijke raadsman. Geduldig maakte hij mij de problemen van het wetenschappelijk werk duidelijk en wees de weg naar oplossingen. Prof. Dr. W.H.L. Hoefnagels wil ik danken voor zijn steun bij de wetenschappelijke, organisatorische en klinisch geriatrie aspecten van het onderzoek, maar vooral voor zijn enthousiasmerende invloed, ook als mijn chef op de afdeling Geriatrie. Prof. Dr. B.C.M. Raes, destijds hoofd consultatie-dienst psychiatrie van het Sint Radboud ziekenhuis, speelde in de beginfase van het onderzoek een voorname rol. Zonder zijn stimulerende invloed was ik er niet eens aan begonnen. Ook zijn hulp bij de behandeling van psychopathologische kwesties is belangrijk geweest. Dr. J.R.M. Cruysberg, mijn contactpersoon met de afdeling oogheelkunde, ben ik dankbaar voor zijn hulp bij de organisatie van het onderzoek, de behandeling van ophthalmologische kwesties en -niet in het minst- zijn esthetisch schaven aan de artikelen. Prof. Dr. A.L.M. Verbeek heeft onontbeerlijke methodologische hulp geboden en mij ruim laten profiteren van zijn ervaring als publicist. Dr. P.J.E. Poels heeft mij bijgestaan als het ging om de neurologische aspecten van het syndroom. Dr. Y. Kuin heeft geholpen geschikte psychologische onderzoeksinstrumenten te vinden en de bevindingen te interpreteren. Dr. M. van 't Hoff bood uitkomst toen statistische kennis op hoog niveau gevraagd werd.

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Ik ben de stichting Theofaan erkentelijk, die mijn deelname aan het Low-Vision congres te Madrid in 1996 mogelijk heeft gemaakt en ook bijgedragen heeft aan het totstandkomen van dit proefschrift.

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Curriculum vitae

De auteur van dit proefschrift werd geboren op 11 mei 1955 te Ermelo. In 1974 behaalde hij het diploma Atheneum-B aan de Rijksscholengemeenschap te Harderwijk. Van 1974 tot 1975 vervulde hij de militaire dienstplicht. Vanaf 1975 studeerde hij geneeskunde aan de Rijksuniversiteit te Utrecht, behaalde het kandidaats-examen cum laude in 1978 en ontving in 1979 de beloning ex artikel 60 W.W.O. (bevordering studiezijn) naar aanleiding van zijn wetenschappelijke stage (onderzoek naar de remming van het effect van β -endorphine op de poetsactiviteit van ratten door herhaalde intraventriculaire toediening; stage-leider: Prof. Dr. W.H. Gispen). In juni 1982 behaalde hij het arts-examen. Van januari 1983 tot mei 1985 was hij als arts in opleiding tot neuroloog verbonden aan de afdeling Neurologie van het Evangelische Johanneskrankenhaus te Bielefeld, Duitsland (opleider: Dr. E. Ebel). Van juni 1985 tot december 1989 volgde hij de opleiding tot psychiater (hoofdopleider Dr. J.W.A. Hubert) met de basisopleiding in het Psychiatrisch Centrum Zon en Schild, de stage sociale psychiatrie bij het RIAGG Westelijk Utrecht (opleider Drs. K.H. Lamberts) en de stage gerontopsychiatrie in het Psychiatrisch Centrum Venray (opleider Drs. J.J.C. Marlet). Sinds januari 1990 is hij als psychiater -met als bijzonder aandachtsveld de ouderenpsychiatrie- verbonden aan de de afdeling Psychiatrie (hoofd: Prof. Dr. F.A.M. Kortmann) van het Academisch Ziekenhuis Nijmegen. Tot maart 1996 is hij tevens gedetacheerd werkzaam geweest op de afdeling Geriatrie (hoofd: Prof. Dr. W.H.L. Hoefnagels) van het zelfde ziekenhuis. Vanaf januari 1998 is hij lid van het bestuur van de Sectie Ouderenpsychiatrie en van de Commissie Wetenschappelijke Activiteiten van de Nederlandse Vereniging voor Psychiatrie.

Rob Teunisse is sinds 1986 gehuwd met Elke Dellbrügge en heeft drie kinderen: Steven (geb.1989), Esther (geb. 1992) en Nico (geb. 1994).

