Low awareness of the Charles Bonnet syndrome in patients attending a retinal clinic

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ABSTRACT

INTRODUCTION: Visually impaired patients may experience visual hallucinations due to the Charles Bonnet syndrome (CBS). While benign in nature, these hallucinations may cause distress in those unfamiliar with the phenomenon. The overall purpose of this study was to determine the degree of awareness of CBS in patients referred to our retina clinic.

MATERIAL AND METHODS: Consenting patients attending our retina clinic over a period of three months underwent a thorough ophthalmological examination, including slit-lamp fundus biomicroscopy, spectral-domain optical coherence tomography, fundus autofluorescence imaging and fluorescein/indocyanine green angiography (if applicable). Visual acuity was measured and the participants were subjected to a structured telephone interview.

RESULTS: A total of 200 patients were included in this cross-sectional study. Twelve percent of the participants were familiar with CBS. Patients who were clients at a low-vision rehabilitation clinic or were highly educated were more likely to be familiar with CBS. There was an association between low visual acuity and awareness of CBS. Logistic regression analysis revealed that only low visual acuity and university education were independently associated with familiarity with CBS. Fifteen percent of the participants admitted to having visual hallucinations.

CONCLUSION: Visually impaired patients are largely unfamiliar with CBS. Since unawareness of CBS may cause unnecessary distress in some patients, efforts to educate low-vision patients about CBS should be made.

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The Charles Bonnet syndrome (CBS) is a benign condition characterised by recurrent visual hallucinations commonly occurring in visually impaired persons. The hallucinations are typically described by patients as being vivid, complex, recurrent and clearly defined [1, 2]. While most patients will identify the contents of their hallucinations as being unreal, about a third may suffer from some to severe distress due to fear of impending insanity [1, 3]. The prevalence of CBS in a Danish population with late age-related macular degeneration (AMD) was found to be 8.3%, and other studies have reported a prevalence ranging from less than 1% to one that reaches 40% [3-10]. Even though a relatively large percentage of low-vision patients are thus experiencing potentially distressful visual hallucinations, it remains unknown how informed these patients are about the relationship between visual deprivation and visual hallucinations. We therefore set out to determine how large a proportion of the patients attending our retina clinic knew of CBS, and whether certain socio-demographic factors were of any importance in this context.

MATERIAL AND METHODS

Study subjects and clinical data

A total of 246 consecutive patients attending our retinal clinic over a three-month period were invited to participate in this cross-sectional study. The participants were either being assessed for treatment response to anti-vascular endothelial growth factor after treatment of exudative AMD or other retinal conditions (retinal vein occlusion or diabetic maculopathy), or they were newly referred patients suspected of having exudative AMD or neovascularisation secondary to other retinal conditions potentially requiring treatment. The best corrected visual acuity (BCVA) was measured in both eyes using the Early Treatment of Diabetic Retinopathy Study (ETDRS) chart and converted into decimals. The retinae were clinically examined with slit-lamp biomicroscopy using a 90-diopter lens. Retinal imaging using spectral-domain optical coherence tomography, fundus autoflourescence imaging and flourescein/indocyanine green angiography (where applicable) was performed in all patients. All patients were asked by the receiving nurse if they would be interested in a short phone interview about their eyesight for a research project. Out of the 246 patients asked in total, 22 refused to participate for unknown reasons. The remaining patients were called on phone by author AS or YS, and 24 did not answer the phone despite several attempts (a minimum of three attempts on different days). On the phone, the patients were asked the following question (translated into Danish): "Some patients with poor eyesight see things which they know are not there. These hallucinations occur on repeated occasions and are described as being vivid and complex, e.g. persons, animals, flowers, or patterns. This phenomenon is known as the Charles Bonnet syndrome. Have you ever heard or read about this?" The patients were

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also asked if they had personally experienced CBS. It was noted whether or not patients had visited a low-vision rehabilitation clinic (BCVA < 0.3 (less than 60 ETDRS letters) on the better eye) for an ocular condition. Finally, we inquired about the patients' highest achieved educational level and usage of the internet for informational purposes only. Participants reporting elementary hallucinations, e.g. photopsias or shadows, were not included in the study. Current and previous ocular diagnoses and a history of psychiatric or neurological disease were noted from the patients' records.

Statistical analysis was performed using SPSS 20 for Windows (IBM, Chicago, IL, USA). Student's independent t-test was applied to normally distributed continuous variables (age, duration of disease) and the average was provided as a mean with standard deviation. χ^2-test or Fisher's exact test (where applicable) were used for categorical variables (male/female ratio, history of CBS, primary ophthalmic diagnosis, pre-existing or current ocular conditions, client at low-vision clinic, psychiatric or neurological conditions, level of education and internet usage). The Mann-Whitney test was used where continuous data were non-parametric (visual acuity) and the average was provided as a median with the interquartile range. A p-value < 0.05 was considered statistically significant.

Trial registration: not relevant.

RESULTS

A total of 200 patients were included in this study (**Table 1**). The mean age of the participants was 78.5 (standard deviation 8.2) years. There were 61 males (30.5%) and 139 (69.5%) females. A total of 30 (15%) patients admitted to having complex visual hallucinations characteristic of CBS. The majority of the patients (91.5%) had AMD, 3.5% had vein occlusion, 1% had diabetic retinopathy and the remaining 4% had various other retinal diagnoses (see Table 1). In all, 12% of participants were familiar with CBS, while the remaining 88% had never heard or read about it. There were no differences in the male/female ratio (p = 0.43, χ^2-test), duration of eye disease (p = 0.14, t-test), primary ophthalmic diagnosis (p = 0.41, χ^2-test), pre-existing or current ocular or neurological/psychiatric conditions (p = 0.26, Fisher's exact test; p = 0.16, χ^2-test, respectively), or internet usage (p = 0.41, χ^2-test) between patients who were familiar with CBS and patients who were unfamiliar with CBS before

TABLE 1

Characteristics of study participants.

	Familiarity with CBS?		
	no (N = 176)	yes (N = 24)	p-value
Age, yrs, mean (± SD)	78.6 (± 8.4)	78.4 (± 6.5)	0.90
Females, %	70.5	62.5	0.43
Duration of disease, months (IQR)[a]	13.5 (1-30)	19 (6.5-48.5)	0.14
History of CBS, %	13.1	29.2	0.061
Primary ophthalmic diagnosis, %		0.41	
AMD	92.6	87.5	
Central or branch vein occlusion	3.4	4.2	
Diabetic maculopathy or retinopathy	0.6	4.2	
Others[b]	3.4	4.2	
Best corrected visual acuity in the better eye (IQR)	0.63 (0.4-1.0)	0.45 (0.2-0.63)	0.014
Pre-existing or previous ocular conditions[c], %	8.5	16.6	0.26
Client at a low-vision clinic, %	17.6	41.6	0.013
Previous or current psychiatric or neurological conditions, %	35.2	20.8	0.16
Level of education, %			0.012
No education	41.5	25.0	
High school/vocational courses	47.2	41.7	
University	11.4	33.3	
Internet usage for informational purposes only, %			0.41
Daily	18.8	33.3	
Weekly	12.5	12.5	
Rarely	6.3	4.2	
Never	62.5	50	

AMD = age-related macular degeneration; CBS = Charles Bonnet syndrome; IQR = interquartile range; SD = standard deviation.

a) Estimated from date of first contact.

b) Included degenerative myopia, central serous chorioretinopathy, pseudovitelliform dystrophy, amblyopia, peripheral exudative haemorrhagic chorioretinopathy, corneal oedema, and subretinal tumour.

c) Minor ocular conditions, such as refractive errors and incipient cataracts were not included.

being asked about these issues. Patients familiar with CBS were more likely to have a history of CBS (29.2% versus 13.1%); this difference was close to reaching statistical significance (p = 0.061, Fisher's exact test). Patients familiar with CBS were more likely to be clients at a low-vision clinic (p = 0.013, Fisher's exact test), and they were more likely to be highly educated (p = 0.012, χ^2-test).

BCVA in the better eye was significantly lower in patients who were familiar with CBS (p = 0.014, Mann-Whitney U test). In a multiple logistic regression analysis with familiarity to CBS as the dependent variable and the three covariates (level of education, visual acuity and low-vision clinic client) as independent variables, only university education (p = 0.002; odds ratio (OR) = 7.67; 95% confidence interval (CI): 2.15-27.32) and lower visual acuity (p = 0.045; OR = 6.02; 95% CI: 1.04-34.48) were independently associated with familiarity with CBS. Attachment to a low-vision clinic (p = 0.12; OR = 2.29; 95% CI: 0.80-6.55) or high-school/vocational education (p = 0.23; OR = 2.0; 95% CI: 0.65-5.99) were not independently associated with CBS familiarity.

DISCUSSION

CBS was described about 250 years ago by the Swiss philosopher, naturalist and biologist Charles Bonnet. In the 1930s, de Morsier labelled these symptoms the Charles Bonnet syndrome [11]. Since then, several case reports and epidemiological studies have described CBS as a more or less common condition occurring in people who are deprived of visible light. While the exact pathogenesis of CBS remains poorly understood, it has been proposed that lack of visual stimuli to the visual association areas of the cerebral cortex can trigger visual hallucinations (the de-afferentation theory) [12, 13]. There is no consensus on the criteria used to diagnose CBS, but the Gold and Rabins criteria are widely used in literature [1, 13]. According to these, CBS can be diagnosed if the following four criteria are fulfilled: 1) formed, complex, persistent/repetitive and stereotyped visual hallucinations; 2) fully or partially retained insight into the unreal nature of the hallucinations; 3) absence of primary or secondary delusions; and 4) absence of hallucinations in other modalities. Note that these criteria do not include visual impairment; however, most studies report an inverse association between visual acuity and the risk of CBS. For instance, Gilmour et al found that CBS was present in 34% in low-vision subjects (based on visual acuity alone) compared with only 2% of normal-vision subjects [14].

In this study, we found that only 12% of our participants were familiar with CBS. This is cause for concern, especially when considering the fact that CBS is relatively common in patients with eye disease, and many of these patients will not take the initiative to share their hallucinatory experiences with others. In fact, we have previously shown that 36% of patients do not share their experiences with others [3]. CBS may cause distress in patients unaware of the benign nature and course of CBS; and reassurance is the mainstay of the management of CBS [1, 15, 16].

The Gentofte lake as seen by a person with normal vision (**A**), and a person with impaired central vision and contrast sensitivity with a relative scotoma and a Charles Bonnet image (**B**). Note that the Charles Bonnet image stands in sharp contrast to the obscure background.

Various factors may contribute to the patients' awareness of CBS. We investigated some factors which we hypothesised could play a role in increasing the patients' awareness of CBS, such as duration of disease, having a positive history of CBS, being a client at a low-vision clinic, level of education and internet usage. Being a client at a low-vision clinic increased the likelihood of awareness, as did a poor BCVA. In Denmark, patients who have a BCVA < 6/18 or a significantly reduced visual field are considered suitable for low-vision clinic referral. However, we found no statistically significant differences between BCVA of patients who were referred to a low-vision clinic and patients who did not meet the requirements for referral (median 0.63, interquartile range (IQR): 0.4-1.0; median 0.63, IQR: 0.32-0.85; Mann-Whitney U test, p = 0.094). Thus, it appears that BCVA itself (and independently of low-vision client status) is associated with an increased awareness of CBS. Patients with higher levels of education were more familiar with CBS than those with lower levels of education or no education. In a logistic regression analysis, only university education and low vision were independently associated with familiarity with CBS. A positive history of CBS was also associated with increased awareness, though this difference did not reach statistical significance. Surprisingly, internet usage did not increase awareness, even though the internet has a large amount of easily accessible information regarding CBS. The duration of disease also did not affect awareness, although it should be noted that we did not have exact data on the duration of disease and therefore used the time since first presentation as a surrogate marker for duration of disease, which, in itself, may not necessarily reflect the duration of visual impairment.

There were some limitations to this study. First of all, all interviews were conducted on the telephone

which may have affected the validity of the answers. Some patients may have had a hard time hearing or apprehending the questions, and trust in the interviewer may be harder to build over the telephone. Some patients may have been in the company of others and this could affected their willingness to answer correctly or precisely. Secondly, selection bias may have occurred as some of the patients refused to participate in the study, and others, who did agree, subsequently failed to answer the phone. Finally, the answers of the patients may have differed depending on which interviewer (AS or YS) was calling.

CONCLUSION

Patients attending our ophthalmology department for miscellaneous retinal conditions were largely unfamiliar with the commonly occurring CBS. This is cause for concern as we believe that lack of awareness of CBS may contribute to the distress experienced by some patients. Patients who were clients at a low-vision clinic were more familiar with CBS, as were patients with higher education or lower visual acuity. This study emphasises the need to better inform patients with vision-affecting eye disease about CBS.

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