ORIGINAL ARTICLE

Prevalence and clinical characteristics of Charles Bonnet syndrome in Madrid, Spain

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Purpose: *Charles Bonnet syndrome (CBS) is a condition characterized by development of visual hallucinations in patients with no cognitive impairment and significant loss of vision mainly caused by age-related macular degeneration (AMD) or glaucoma.*
Methods: *This was a study of prevalence and characteristics of CBS diagnosed at the Neuroophthalmic Unit within the Ophthalmology Department of Hospital Clínico San Carlos (HCSC), Madrid, Spain.*
Results: *The CBS prevalence in patients from HCSC Madrid is 0.47%, rising to 15% in patients with low vision. Women over 80 years of age comprised 58.3% of the patients, who mainly had AMD (58.3%). Main characteristics of hallucinations included animals (50%), color (58.3%), moving (75%), 6- to 12-month evolution (50%), three times a day frequency (75%), and 3- to 5-minute duration (50%).*
Conclusions: *Charles Bonnet syndrome is a complex process that must be treated jointly by ophthalmologists, neurologists, and psychiatrists in order to ensure accurate diagnosis and adequate management. New studies are needed in order to improve awareness of clinical manifestation of this condition, the incidence of which is underestimated due to patients' fear of being branded mentally ill, as well as physicians' lack of knowledge about CBS.*

Keywords: *Age-related macular degeneration, Charles Bonnet syndrome, Glaucoma, Low vision, Visual hallucinations*

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INTRODUCTION

Charles Bonnet syndrome (CBS) is a condition characterized by the manifestation of visual hallucinations in patients with severe visual deterioration and no cognitive impairment (1-3). Increasing life expectancy among patients, many of whom show significant visual loss secondary to age-related macular degeneration (AMD) or glaucoma, is causing an increase in the incidence of CBS. Estimated prevalence of simple visual hallucinations in patients with severe visual impairment is 59%, decreasing to 15% for complex visual hallucinations. Despite this, case studies have shown a prevalence of between 1.84% and 3.15%. This is due to lack of awareness on the part of doctors, as well as patients' fear of being diagnosed as mentally ill (1-3). There is great variability among published data and CBS has not been solely described in visually impaired patients of advanced age. Cases have been described in patients with good vision (4), young patients with low vision (5, 6), and children (7, 8).

METHODS

This article surveys CBS prevalence at the Ophthalmology Unit of Hospital Clínico San Carlos (HCSC) in Madrid, Spain. A group of 2502 patients who had previously been examined at the outpatient unit were considered in the

study. They also underwent a comprehensive eye examination that included visual acuity (VA) measurement, anterior pole biomicroscopy, posterior pole examination, intraocular pressure measurement, and study of extrinsic and intrinsic ocular motility. All the patients were asked about the presence of simple and complex visual hallucinations. Those patients with visual hallucinations were referred to the Neuroophthalmic Multidisciplinary Unit—formed by the ophthalmology, neurology, and psychiatry units at HSCS—for further study. Diagnosis of CBS was by exclusion, once other conditions that can trigger hallucinations had been ruled out. Distribution by sex, underlying ocular pathology, type of hallucination, evolution period, frequency, and duration were also analyzed.

RESULTS

Prevalence of CBS varies between 1.84% and 3.15% according to published data. Our study shows a prevalence of 0.47%, rising to 15% when limited to patients with low vision. Most of the patients were women (58.3%) over 80 years of age (58.3%) and had severe visual impairment. All 12 cases had VA below 0.1, mainly caused by AMD (58.3%) (Tab. I). Hallucinations were mainly of animals (50%), in color (58.3%), and moving (75%). Half of the patients (50%) had been experiencing hallucinations for a year, having 3 or more episodes a day (75%), which would last between 3 and 5 minutes (50%) (Tab. II).

TABLE I - POPULATION DISTRIBUTION, VISUAL ACUITY, AND ASSOCIATED OCULAR PATHOLOGY

Characteristics	Values, n (%)
Age, y	
≤70	0
71-80	5 (41.6)
>80	7 (58.3)
Sex	
Female	7 (58.3)
Male	5 (41.6)
Visual acuity	
≤ Count fingers 1 m	4 (33.3)
Count fingers 1 m ≤0.1	8 (66.6)
Associated ocular pathology	
Age-related macular degeneration	7 (58.3)
Glaucoma	3 (25)
Cataract	1 (8.3)
Retinal detachment	1 (8.3)
Other associated pathology	
Hypoacusis	5 (41.6)
Parkinson disease	1 (8.3)
Depression	6 (50)
CBS prevalence in general population	12/2502 (0.47)
CBS prevalence in low vision patients	12/80 (15)

CBS = Charles Bonnet syndrome.

DISCUSSION

Charles Bonnet syndrome is characterized by the presence of visual hallucinations ranging from simple ones such as lines and basic geometric shapes to complex and structured ones usually in the shape of people, faces, animals, or trees, though they do not include sound. They can be black and white or in color, static or moving (9). They last under 10 minutes in most cases and are usually repetitive. Clinical course may be episodic, cyclic, or chronic, with duration usually under 12 months, though some cases lasting several years have been described.

Although the cause behind the manifestation of hallucinations remains undetermined (10), the deafferentation theory would be responsible for the onset of CBS. According to this theory, reduced or absent stimulation of retinal nerve cells leads to decreased stimulation of the occipital cortex, though not disappearing completely as in amaurosis. Residual afferents would then trigger the deafferentation phenomenon through histologic, biochemical, and anatomical changes to synapses in order to try to compensate for the limited stimulation, thus becoming hyperexcitable (10). Onset of CBS has been associated with specific triggering factors such as fatigue, stress, low lighting, and dazzling lights (10). Charles Bonnet syndrome has also been linked to social isolation, cognitive defects, sensory deprivation, and low-quality social interaction (11, 12). Hallucinations were not triggered by any external factors in 58.3% of the patients in our study. Furthermore, the same percentage of patients, 58.3%, did not show any other associated factors such as social isolation or sensory deprivation (Tab. II).

Patients with CBS do not show any cognitive deficiency or any other type of sensory hallucination because it is not a

TABLE II - FEATURES OF HALLUCINATIONS AND TRIGGERING AND ASSOCIATED FACTORS IN CHARLES BONNET SYNDROME

Characteristics	Values, n (%)
Characteristics of hallucinations	
Color	7 (58.3)
Black and white	5 (41.6)
Type of hallucination	
People	1 (8.3)
Plants/trees	4 (33.3)
Animals	6 (50)
Other	1 (8.3)
Movement	
Moving	9 (75)
Static	3 (25)
Development period, mo	
≤1	1 (8.3)
1-3	2 (16.7)
3-6	3 (25)
6-12	6 (50)
Frequency, episodes/d	
1	3 (25)
3	9 (75)
Duration, min	
≤1	1 (8.3)
1-2	1 (8.3)
3-5	6 (50)
6-10	1 (8.3)
11-15	3 (25)
Triggers	
None	7 (58.3)
Dazzling lights	3 (25)
Low light	2 (16.7)
Associated factors	
None	7 (58.3)
Social isolation	2 (16.7)
Sensory deprivation	3 (25)

psychiatric disorder. Noncoexistence with any other sensory hallucination is crucial, since it would imply a psychiatric pathology otherwise. However, CBS plus has been described in patients with vision impairment and hypoacusis. These patients describe hearing musical sounds that they recognize as unreal (13).

Visual loss and advanced age are the 2 main triggering factors of this condition. Visual loss is mainly caused by AMD, cataract, and glaucoma. The main associated pathologies in our study are AMD (58.3%) and glaucoma (25%) (Tab. I). Charles Bonnet syndrome has also been associated with ocular treatments such as photodynamic (14) or antiangiogenic (15) therapies; treatment with topical drugs such as brimonidine (16) and systemic drugs (17); neurosurgery (18); and systemic pathology (19). Charles Bonnet syndrome has also been described in patients with good vision (4) and glaucoma (20), the latter possibly related to deafferentation of injured fibers even if central vision is preserved. Development of CBS may be linked to onset of dementia, a risk that may be up to 20 times higher than in the rest of the population (21).

Treatment of CBS must be comprehensive, must be individually tailored, and must cover all of those aspects related to the etiopathology of the syndrome. Providing patients and their families with adequate information, improving their quality of life, treating the cause if possible, or treating secondary anxiety disorder are all critical in achieving comprehensive treatment. Regarding medical treatment, there is a wide range of therapeutic options that cover all aspects related to the pathogenesis of CBS (22, 23).

In conclusion, prevalence of CBS at HCSC is 0.47%, rising to 15% among patients with low vision. New studies on CBS are needed in order to understand all of the variables associated with this complex syndrome. Collaborative work in a multidisciplinary unit encompassing the fields of ophthalmology, neurology, and psychiatry has proved to be more efficient as it encompasses all the characteristics and aspects associated with CBS.

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