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Charles Bonnet Syndrome: Challenges in recognition and the need to raise awareness in clinical practice

Síndrome de Charles Bonnet: desafios no reconhecimento e necessidade de conscientização na prática clínica

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The world's population is getting older. Increased survival is accompanied by debilitating diseases, both systemic and ocular, due to a wide variety of causes, including cataracts, age-related macular degeneration (AMD), diabetic retinopathy, glaucoma, trauma, and central retinal artery occlusion, among others. There are currently around 45 million blind people and 2.2 billion visually impaired people worldwide[1,2]. Visual impairment not only has personal, family, and financial consequences but can also be accompanied by a condition often unknown to health professionals, called Charles Bonnet syndrome (CBS).

CBS is characterized by the manifestation of visual hallucinations in psychologically healthy individuals who have significant visual impairment as a result of ophthalmic diseases[3]. It is benign in nature but causes great anxiety for patients who, aware that the hallucinations are not real, are often misdiagnosed as having psychiatric disorders or dementia[4].

The term CBS was first introduced by De Morsier in 1967 to characterize this condition of visual hallucinations in elderly patients with intact brain function[5]. The term refers to the observations of the Swiss philosopher Charles Bonnet (1720-1792) who, in 1760, reported the hallucinatory findings perceived by his 89-year-old grandfather, Charles Lullin. Due to advanced cataracts, Charles Lullin had poor visual acuity and reported visions of men, women, birds, and buildings that changed shape, size, and place, but he was aware that these visions were not real. Years later, Charles Bonnet himself had hallucinations similar to those of his grandfather as a result of low vision secondary to cataracts[3,5,6].

Visual hallucination can occur in various clinical situations and is defined as a visual perception in the absence of external stimuli. It can be simple, composed of abstract images, flashes of light, lines, and geometric shapes, or complex, presenting as images of people, animals, plants, or objects[3-7]. The hallucinations can last from seconds to days and may occur daily, monthly, or in episodes throughout the year. The frequency is variable and may be episodic, periodic,

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or continuous. Episodic hallucinations usually occur in isolation, last from days to months, and usually resolve completely. Continuous hallucinations are persistent and do not disappear[3,5,6]. It is not well established whether there are triggers such as fatigue, stress, or lighting levels, but in very disturbed patients, simply closing and opening the eyes can cause the hallucination to disappear[3,6].

The theory proposed for CBS is the phantom limb pain theory, in which pain occurs even when the limb has been removed. Similarly, patients can have visual sensations despite being unable to see. The etiology of CBS is diverse. Apart from congenital blindness, CBS can result from any condition that causes visual impairment or loss, or that affects the visual pathway from the eyes to the visual cortex[4-9]. This includes ocular conditions such as cataracts, corneal opacity, age-related macular degeneration (AMD), glaucoma, retinitis pigmentosa, diabetic retinopathy, optic neuritis, temporal arteritis, venous occlusion, high myopia, and also, neuronal conditions as cerebral infarction, or stroke involving the occipital lobe[5,6,10].

The most widely accepted hypothesis to explain the pathophysiological mechanism of the visual hallucination process in CBS is deafferentation, which is the loss of afferent neurons responsible for vision, usually due to a lesion in the visual pathway. This loss causes hyperexcitability in the visual cortex, resulting in the formation of images, in this case, visual hallucinations. The factors that contribute to this phenomenon include an increase in the release of pre-synaptic neurotransmitters, an increase in the number of post-synaptic receptors, and a decrease in the release of inhibitory neurotransmitters[4,8,9,11-14]. The diagnostic criteria for CBS are also controversial,and include: I) Presence of complex, stereotyped, persistent, or repetitive visual hallucinations; II) Maintained awareness of the unreal nature of the hallucination; III) Absence of hallucinations in other sensory modalities; and IV) Absence of cognitive impairment. All these criteria are necessary for the diagnosis of CBS[8,11,15].

CBS mainly affects elderly patients, but it can occur at any age. According to the reports in the literature, the average age of patients varies between 70 and 80 years3-12.Some studies have reported the syndrome in younger patients, aged 29-52, with no gender predilection[3,8,11-14]. According to the studies, the prevalence ranges from 0.4% to 36%, a percentage that is underestimated due to the high number of underdiagnoses, whether because patients are unaware of the condition or because they are afraid to report their hallucinations for fear of being stigmatized as having a psychiatric disorder or mentally incapacity[16,17]. In the 1990s, studies considered CBS to be relatively common through an analysis of prevalent studies in the period, and it is estimated that one in seven elderly patients have had a hallucinatory experience, with simple visual hallucinations being more common than complex ones[15,16].

Therefore, a lack of knowledge and understanding of CBS can lead to misdiagnoses of dementia, Alzheimer's, and schizophrenia. These misdiagnoses can result in patients receiving incorrect treatments, which can have a negative impact on their quality of life. This impact can stem from the emotional stress caused by the fear of having dementia or a psychiatric illness, as well as from the failure of the therapies provided to alleviate hallucinations[8,9,11,12,16,17].

Currently, there is no effective therapy for CBS. As life expectancy increases and the population ages, it is crucial for health professionals to recognize this condition to ensure accurate diagnosis and provide proper guidance to patients and their families. Recognizing CBS is essential for reducing anxiety, offering appropriate care, and fostering a healthy family dynamic, all of which positively impact the quality of life for everyone involved.

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