



Charles Bonnet syndrome: a condition of the visually impaired

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The number of low vision patients that suffer from visual hallucinations is shockingly large. The systematic review and meta-analysis by Subhi *et al.* in this edition of *AES* shows that the prevalence of Charles Bonnet syndrome (CBS) in the low vision population is approximately one in five. CBS is defined by the World Health Organization (WHO) as ‘the experience of complex visual hallucinations in a person who has experienced partial or complete loss of vision’ (1). The hallucinations are exclusively visual, usually temporary, and not associated with mental and behavioral disorders. The hallucinations negatively affect daily activities and result in anxiety and stress in as many as one third of CBS patients (2-4). Understanding the phenomenon of CBS by these patients can alleviate these problems, however, most CBS patients are reluctant to share their hallucinogenic experiences out of fear of being labelled mentally unstable. Conversely, clinicians do not routinely check for these symptoms, as a result, CBS is often overlooked in ophthalmic practice (5).

Subhi *et al.* identified 11 studies that systematically analysed patients for the presence of CBS. Their meta-analysis showed a pooled prevalence of CBS in low vision patients of 19.7% (95% CI: 13.8–26.4%). Extrapolating this prevalence results in an estimate of 47 million people worldwide that experience CBS with roughly one-third (16 million) experiencing fear and stress as a result. The first step in addressing CBS is the recognition by ophthalmologists that this condition is highly prevalent in our patient population.

The risk of CBS seems to correlate with the extent of vision loss, rather than the underlying ophthalmic diagnosis. In addition, recent loss of visual acuity also seems to be a risk factor for the development of CBS (6). Subhi *et al.* report various diseases along the visual pathway that result in CBS including cataract, age related macular degeneration (AMD), and glaucoma. In addition, neurological diseases that cause a visual field defect may also result in CBS (7). Subhi *et al.* found the prevalence of CBS in patients with cataract to be similar to CBS prevalence in reports that focused on retinal diseases. We should therefore actively identify patients with visual hallucinations among all patients with moderate to severe visual impairment (visual acuity lower than 6/18, but higher than 3/60), regardless of the underlying diagnosis, be it ophthalmic or neurologic.

Another misconception is that CBS is age-related and primarily affects the elderly low vision patient (8-13). This assumption probably arises from the fact that most CBS studies were carried out in patients with age-related diseases like AMD, glaucoma and cataract (3,4,14). However, there is a growing body of more recent literature that shows that younger age groups are also affected and even paediatric cases have been reported (15-18). Unfortunately, the meta-analysis of Subhi *et al.* only included adults aged ≥40 years. Yet, none of the included studies could prove a statistically significant difference in the prevalence of CBS across different age groups. Our group recently reported a CBS prevalence of 8.4% in a cohort of 83 relatively young patients with Stargardt disease (6). This relatively high

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prevalence suggests that CBS is probably underestimated in younger patient groups, stressing the need for active identification of these patients.

A major difficulty in studying the collective data on CBS lies within the variable definition, or lack thereof, of this syndrome. Indeed, in the meta-analysis of Subhi *et al.*, a clear definition of CBS was absent in six out of the 11 studies. Over the years, various diagnostic criteria for CBS have been proposed. This leads to the peculiar notion that investigators even disagree as to the significance of decreased vision as a criterion for CBS. The majority of information on CBS is reported in psychiatric and neurologic rather than ophthalmic literature. Neurologist De Morsier, who coined the term CBS, and psychiatrists Gold and Rabins, who developed a commonly used CBS definition, consider visual impairment not obligatory for diagnosis (19,20). The criteria as proposed by Teunisse *et al.* also do not include visual impairment as a vital component of the syndrome, when in fact they found a clear association between visual impairment and CBS (8,21). Podoll *et al.* are one of the few who have included loss of vision as a consequence of ocular disease in their criteria as a specifying factor (22). In contrast, all ophthalmologists who have described CBS, consider visual impairment a main criterion in the diagnosis of CBS (7).

In the 2018 revision of the International Classification of Diseases, the WHO has allocated a specific diagnose code for CBS (9D56) (1). Based on this WHO definition, the principal diagnostic criteria include: (I) the presence of complex visual hallucinations; (II) partial or complete loss of vision; (III) the absence of mental or behavioural disorders. The pooled prevalence of almost 20% that was reported by Subhi *et al.* calls for systematic inquiry for presence of CBS among the visually impaired. We therefore would like to challenge you as an ophthalmologist. Pose the following question to your patients with a vision impairment in the range of 3/60-6/18: 'Have you ever seen things or images that you know were not actually present at that moment?'. The number of patients that may answer your question with a 'yes' may surprise you.

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