

# CHARLES BONNET SYNDROME

We explore an underdiagnosed complication of diabetic retinopathy

## What is Charles Bonnet syndrome (CBS)?

Charles Bonnet syndrome (CBS) is named after the Swiss philosopher Charles Bonnet, whose father experienced visual hallucinations after losing his sight following cataract surgery in the 18<sup>th</sup> century. People with CBS experience hallucinations triggered by loss of vision due to an eye condition, for example diabetic retinopathy, age-related macular degeneration, cataracts or glaucoma. These tend to start within weeks or months of visual loss and don't involve hearing or any other sensations. Most hallucinations last seconds or minutes, though can last longer and over time may become shorter. Commonly reported forms of hallucination include colours and shapes, geometrical patterns, distorted faces, and costumed figures<sup>1</sup>.

CBS risk increases in patients with severe impairment of visual acuity and a key risk factor is acuity of 6/36 or less. Patients are aware that what they are seeing is not real<sup>2</sup>.

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## What causes CBS?

If the retina is damaged, the stream of electrical impulses to the visual cortex is reduced. It is believed that in CBS, the brain cells in the visual cortex responds paradoxically by firing more signals, causing visual hallucinations, and studies have shown cortical hyperexcitability<sup>3,19</sup>.

## How is CBS diagnosed?

CBS is a diagnosis of exclusion. Other conditions that can cause transient complex visual hallucinations and must be ruled out include hypnagogic hallucinations, epileptic phenomenon, Parkinson's disease, Lewy body dementia, and migraine. Persistent complex visual hallucinations can also be caused by Anton's syndrome, thalamic or brain stem lesions and delirium tremens. CBS can be diagnosed when these conditions have been ruled out in someone with vision

loss and visual hallucinations, and in the absence of psychosis or delirium.

CBS is diagnosed via history taking and discussing symptoms with the person, as well as carrying out tests to rule out other possible causes.

## What is the prevalence of CBS?

The condition can occur in any age and it has been described in young children<sup>4</sup>. However, it is more common in the elderly because of the mean age of sight-threatening eye diseases, including age-related macular degeneration, diabetic retinopathy and glaucoma. It's difficult to know how many people in the UK have CBS because quite often people don't always want to tell doctors that they are experiencing the condition. Far too many people who develop CBS have received no warning about the condition and, consequently, confide in no one, according to the NHS<sup>20</sup>.

Despite the distressing nature of CBS, visual hallucinations are seldom

addressed during routine clinical assessments. Patients rarely volunteer information about hallucinations due to fears of being diagnosed with a psychiatric condition or cognitive impairment, and lack of awareness among healthcare professionals<sup>5</sup>. This is supported by a clinical survey highlighting fear of stigma preventing people self-reporting their symptoms<sup>6</sup>.

Depending on the study, the reported prevalence ranges from 0.4% to 30% among those with sight loss<sup>7</sup>. It's thought there are more than 100,000 cases in the UK<sup>20</sup>.

## How long does the condition last?

The widely held belief is that CBS reduces over time and previous longitudinal studies suggest 28% recovery at one year<sup>8</sup> or an average duration of 18 months<sup>9</sup>. This is now, however, being challenged. There is

emerging evidence that CBS may not be as short-lived as previously thought. One study has reported hallucinations continuing for four years or more in 45% of people with CBS<sup>10</sup> but more than 75% of patients will continue to experience hallucinations beyond five years after their onset<sup>11</sup>.

Another found 41% of people with CBS had an estimated minimal average duration of eight years (minimal as hallucinations were ongoing at the end of the study)<sup>12</sup>. It is difficult to dismiss CBS as a clinical irrelevance if it results in troubling symptoms that persist for many years for a large group of people.

## The impact of CBS

Patients' experience of their hallucinations varies widely, from fear to indifference and even to enjoyment<sup>13</sup>. Though some reporting suggests that most patients with CBS do not find it troublesome, around a third find the hallucinations an unpleasant experience<sup>1,6,8,17</sup>. Compared to patients with the same degree of visual loss without hallucinations, patients with CBS have decreased measures of quality of life and functional ability<sup>15</sup>. A third of patients find it interferes in their day-to-day life and it can make moving around and navigating difficult and affect confidence. Around one-third report negative sequelae, including stress linked to uncertainty over the origin and meaning of the hallucinations and if patients are not told about the possibility of CBS with sight loss, this has been shown to increase stress<sup>13</sup>. Distress is increased, too, when trying to cope with vision loss.

## Treatment

There is currently a lack of evidence as to how to treat hallucinations and The National Institute for Health Research funded a five-year research programme resulting in consensus guidelines for their clinical management<sup>16</sup>. Education about CBS, including telling patients about the possibility of hallucinations, is the mainstay of current treatment and sensitive inquiry as to the presence of visual hallucinations is considered an important part of the clinical consultation in at-risk patient groups.

Patients benefit from healthcare professionals being aware of CBS and signposting them to resources that can help. Due to the stigma concerning mental health, reassuring patients that they do not have a mental health problem can help. For most patients, this response will be sufficient, however

support should be offered to those who cannot cope with their hallucinations<sup>17</sup> and referral to specialist services may be required. It has been recognised that support and advice for the carer of the person with hallucinations should be provided too<sup>16</sup>.

### Techniques for minimising or eliminating visual hallucinations secondary to Charles Bonnet syndrome<sup>12</sup>

- When the hallucinations start, look from right to left once every 15 seconds without moving your head.
- Try to touch the hallucination.
- Stare straight at the hallucination.
- Turn your head to alternate sides, then move the head towards each shoulder in turn.
- Walk around the room or to another room.
- Shine a torch from below your chin in front of (not into) your eyes.
- Change the light level in your room or the activity you are doing.

### Impact of Covid-19

A survey conducted between June and July 2020 found that half of respondents experienced exacerbation of visual hallucinations during the Covid-19 pandemic, including increased frequency and more problematic visual hallucinations. Almost half of respondents believed a reduction in physical activity was contributing to these changes. Inactivity is a known risk factor in CBS and the authors of the study recommended support to increase activity levels, through home exercise programmes.

Just under a third of respondents attributed negative or upsetting news stories to increased frequency and onset of more distressing hallucinations. This may partly be explained by loneliness or environmental triggers. Further recommendations to help symptoms in CBS included increasing social interactions, activity and reducing news exposure. It was also noted that healthcare professionals should be aware of CBS risk factors and be knowledgeable about strategies to help patients self-manage CBS<sup>18</sup>.



### Information and support

- Esme's Umbrella includes a section for healthcare professionals listing research and journal papers. Visit [charlesbonnetsyndrome.uk](http://charlesbonnetsyndrome.uk)
- RNIB offers information, support and eye health team that answers calls on the Esme's Umbrella helpline on 020 7391 3299.

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