The Charles Bonnet syndrome: Description in search of etiology

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The Charles Bonnet syndrome is an eponym first proposed by de Morsier in 1938 to name a cluster of symptoms originally described by Swiss naturalist Charles Bonnet in 1760. In his published account, Bonnet reported that his grandfather, who suffered from dense cataracts and had a "sane and reasonable mind," experienced visual hallucinations. In 1967, de Morsier reviewed 18 case reports of this syndrome in the medical literature, concluding that visual hallucinations may occur in individuals without overt neuropsychiatric disease and without obvious peripheral optic disease.

During the past two centuries, the literature reporting the Charles Bonnet syndrome has been largely descriptive and sporadic. It has been confined — with few exceptions — to single- and double-case reports. Although a meta-analysis of these case reports is not the object of this paper, certain symptom similarities have appeared that make the clinical syndrome worthy of further study. Table I outlines the criteria used by investigators from different clinical settings to establish some diagnostic consistency. In Table II, we have culled relevant works in this area to define uniform diagnostic criteria. They are cited here in a composite form so we may continue to use a consensus criteria prospectively to search for a neuropsychophysiologic basis and investigate possible prognostic value for this syndrome.

Does the Charles Bonnet syndrome exist? The reported accounts of the Charles Bonnet syndrome differ considerably. Often, the syndrome has been defined as visual hallucination of formed imagery in the elderly, whose mental status and sensorium are clear. The individuals are reported to be without psychiatric or neurological history. Ocular pathology, for some investigators, has been an essential ingredient. Although initially described in individuals with cataracts and subsequent vision loss, the syndrome has been described in people with any ophthalmological dysfunction: the eye and peripheral optic apparatus, chiasmal, post-chiasmal and any disease of various causes along the optic tracts from the chiasm to the occiput, including the visual association areas. The syndrome also has been reported in individuals without any significant ophthalmologic pathology.

A hallmark of the Charles Bonnet syndrome is that the diagnosis, for most investigators, disallows hallucinations in other sensory modalities. However, the literature is not consistent. Muscular hallucinations have been described concomitantly. Although the visual hallucinations typically do not speak to the affected individual, this finding is not uniform. Tactile and olfactory hallucinations also have been described.

As a diagnosis made with little consideration to clinical course or pathophysiological basis, the Charles Bonnet syndrome has had limited descriptive value. It has been a variable diagnosis of exclusion. In order to make the diagnosis, primary causes of visual hallucinations from psychiatric, neurologic, metabolic, pharmacologic and/or toxic disorders have been omitted. The causes for visual hallucinations, identified by the authors in a separate publication, often include central nervous system disease from tumor or degenerative, demyelinating, ischemic,
**Table 1 — Criteria and clinical features of the Charles Bonnet syndrome**

<table>
<thead>
<tr>
<th></th>
<th>de Morsteker 1967&lt;sup&gt;1&lt;/sup&gt;</th>
<th>Damas-Mora, et al, 1982&lt;sup&gt;2&lt;/sup&gt;</th>
<th>Gold and Rabins 1989&lt;sup&gt;4&lt;/sup&gt;</th>
<th>Siarkowski 1990&lt;sup&gt;9&lt;/sup&gt;</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>ONSET</strong></td>
<td>Variable</td>
<td>Sudden</td>
<td>—</td>
<td>Sudden</td>
</tr>
<tr>
<td><strong>DURATION</strong></td>
<td>Variable</td>
<td>Variable</td>
<td>Persistent</td>
<td>Variable</td>
</tr>
<tr>
<td><strong>LOCALIZATION</strong></td>
<td>Subject dependent</td>
<td>External space</td>
<td>Variable</td>
<td></td>
</tr>
<tr>
<td><strong>CHARACTERISTICS OF VISION</strong></td>
<td>Micro-, macroscopic changing or mobile; vivid and animated</td>
<td>Vivid, human figures, animals, flowers, animated and multicolored</td>
<td>Formed, complex, stereotyped</td>
<td>Organized, defined, detailed and dynamic</td>
</tr>
<tr>
<td><strong>EMOTIONAL REACTION</strong></td>
<td>Variable</td>
<td>Variable</td>
<td>Variable</td>
<td>Variable</td>
</tr>
<tr>
<td><strong>INSIGHT AND COGNITION</strong></td>
<td>Retained</td>
<td>Retained</td>
<td>Usually retained</td>
<td>Retained</td>
</tr>
<tr>
<td><strong>SEX</strong></td>
<td>Predominantly male</td>
<td>Female</td>
<td>Predominantly female</td>
<td>Predominantly male</td>
</tr>
<tr>
<td><strong>AGE</strong></td>
<td>Elderly</td>
<td>Elderly</td>
<td>Elderly</td>
<td>Elderly</td>
</tr>
<tr>
<td><strong>SECONDARY DELUSIONS</strong></td>
<td>Absent</td>
<td>Absent</td>
<td>Absent</td>
<td>Absent</td>
</tr>
<tr>
<td><strong>HALLUCINATIONS IN OTHER MODALITIES</strong></td>
<td>Absent</td>
<td>Absent</td>
<td>Absent</td>
<td>Absent</td>
</tr>
<tr>
<td><strong>OCULAR PATHOLOGY</strong></td>
<td>Unimportant</td>
<td>May be associated</td>
<td>Not necessary</td>
<td>Necessary</td>
</tr>
<tr>
<td><strong>EFFECT OF EYE CLOSURE</strong></td>
<td>Independent</td>
<td>Variable</td>
<td>—</td>
<td>Visions disappear</td>
</tr>
<tr>
<td><strong>NEUROPSYCHO-PATHOLOGY</strong></td>
<td>Undetermined</td>
<td>Undetermined</td>
<td>May be associated with Dementia, delirium, frontal ischemia</td>
<td>Undetermined</td>
</tr>
</tbody>
</table>

Inflammatory, irritative, migrainous, infectious and endocrinological dysfunction. Confusional states have been excluded as etiological, as have electrolyte disturbances, major mental illness, psychological trauma, cultural rituals, hallucinations of widowhood and hostages, hypnagogy and hypnopompic hallucinations, childhood imagery, drug use, abuse and withdrawal, hearing impairments and aging.<sup>4,18,19</sup> Unfortunately, exclusionary criteria have changed as new publications have emerged.

Exclusions aside, the Charles Bonnet syndrome has had other associative features described. It has been mentioned in individuals with delusional beliefs, paranoia, forgetfulness, loneliness and confusion.<sup>11,15,20</sup> Visual hallucinations have been reported subject to alterations in position of the patient, day-night variation, changes in lighting, changes in interpersonal settings, changes with eye closure, visual acuity or visual field disturbances and secondary to bereavement.<sup>4,6,21</sup> It has been described more often in women and/or certain groups of creative or ingenious people such as artists, botanists and university professors.<sup>4,18,22</sup>
Two important features result from the criteria set forth in tables I and II and the myriad of associative findings. If the Charles Bonnet syndrome is to have any meaning, diagnostic accuracy must be clear and useful. The Charles Bonnet syndrome must be differentiated from organic hallucinosis of other cause. Associative findings may blur the differences. Although the syndrome, whose etiology is likely multifactorial, is diagnosed as a symptom cluster at a point-in-time, the neurophysiologic basis and the ophthalmological findings must be critically examined. At the same time, longitudinal studies, infrequently undertaken thus far, must examine whether the syndrome has prognostic significance.

Hypothetical Implications
Based on very few cases, several different etiologies have been proposed to explain this syndrome. The ophthalmological explanation advances the view that, peripherally or centrally, ocular dysfunction is causal. Psychological theory bases the premise of conversion of early life images, memory traces and wish fulfillment into visual hallucinations that then serve a compensatory purpose.13,19,23 Localization theories — which suggest that the cerebral origin of visual hallucinations discriminate according to type, form and color — have not been well substantiated.24,12,36,27 The widely held belief that temporal lobe foci exclusively produce formed images and occipital lobe foci produce unformed images has been questioned seriously. Irritative lesions (epileptiform), migrainous, tumor and other central nervous system pathology (arteritis, ischemia) have been implicated in the localization explanation without reproducibility.6,18,28,29 The “release” explanation — proposed to explain this syndrome in the context of blindness, sensory deprivation or dim light — suggests that the lack of visual afferent input destablizes ophthalmologic processes that, when disinhibited, use stored visual traces to produce imagery.12,13,24,30 Unfortunately, with each of these explanations, the supporting evidence is minimal.

| Table II — Composite criteria for the possible diagnosis of Charles Bonnet syndrome |
|-----------------|----------------------------------------------------------------------------------|
| 1               | Prominent visual hallucinations with multiple forms and colors, including people, plants and animals; imagery may be in black and white |
| 2               | Visual imagery generally occurs in individuals older than 65 with clear sensorium and without obvious neuropsychopathology or pharmacological cause |
| 3               | Images may be identified as unreal |
| 4               | Hallucinosis does not occur in other sensory modalities |
| 5               | Concurrent ophthalmologic pathology, central or peripheral |

Questions
For this syndrome to be clinically useful, certain questions need to be asked and relevant data evaluated:
- If ocular disease is causal, particularly macular degeneration and/or cataract formation, why do the elderly — who are at greatest risk for these ophthalmologic changes — not reveal more cases?
- If isolation, monotony, sensory deprivation or bereavement may affect visual hallucinosis, why are so few elderly apparently affected or reported?
- If visual deprivation is a precursor to visual hallucinosis, should not hearing deficits and auditory hallucinosis be reported with similar incidence in the same population?
- What happens to individuals who have been diagnosed with Charles Bonnet syndrome? Although there are a few cases observed over several years, most reports are of cases briefly studied. Follow-up inquiry is mandatory.
- Quantifiable investigations are rare. Statements such as “his mental status was good for his age” are useless. Prospective studies need careful neuropsychological testing, neuropsychiatric and neurologic assessment, ophthalmological examina-
tion, visual evoked potentials, EEG and quantitative EEG, brain neuroimaging with MRI and cerebral metabolic studies with single photon emission computerized tomography (SPECT) or photon emission tomography.

What parameters define a likely multifactorial etiology?

A prospective view
The authors have begun to examine some of these questions in a prospective manner. Eight cases have been identified that meet our criteria for the Charles Bonnet syndrome. After more than 48 months of observation, the cases demonstrate some striking findings. Ranging from 39 to 88 (seven of the eight are older than 74), six men and two women have been studied. Five of the six men developed Alzheimer’s-type dementia. One of the women developed multi-infarct dementia. Neither of the other two persons developed dementia, although one of the two women died one year after the diagnosis of Charles Bonnet syndrome.

These preliminary results may prove particularly interesting in response to a 1986 study by Hinton et al. on optic nerve degeneration in Alzheimer’s disease. We plan to continue our longitudinal study with further anatomical examination of the optic nerve via MRI, cerebral metabolic studies with SPECT and electrical studies via visual evoked potentials. This may reveal insights into a syndrome that, as yet, eludes a neuropathophysiologic basis.

References


