Charles Bonnet’s Syndrome

Dr Kathryn Bundle, Mayur Bodani, and Lal Landham, in ACNR, June 2007 described Pregabalin in the treatment of visual hallucinations in Charles Bonnet Syndrome (CBS). Their patient had optic atrophy secondary to unrelieved papilloedema with no light perception in the left and minimal light perception in the right eye. The background to Bonnet’s syndrome and its putative explanation may therefore be of interest.

Charles Bonnet (1720-93) (Figure 1) was a Swiss lawyer whose main recreations were as a naturalist and philosopher. In 1760 he described vivid, complex visual hallucinations in his psychologically normal 87-year-old grandfather, Charles Lullin, who had had cataract operations on both eyes and was practically blind (Figure 2). Lullin described silent visions of men, women, birds, carriages, and buildings, which he fully realised were ‘fictions’ of his brain. Bonnet himself later underwent visual deterioration and experienced hallucinations typical of the syndrome named after him by de Morsier in 1936. In this original paper he defined the Charles Bonnet syndrome:

“Dans les syndromes séniiles avec lésions oculaires—le syndrome de Charles Bonnet—(les hallucinations visuelles) peuvent être isolées avec intégrité complète des autres fonctions cérébrales”.

“In the senile syndromes with ocular lesions the syndrome of Charles Bonnet- (visual hallucinations) can be isolated with complete integrity from other cerebral functions.”

He recognised that visual hallucinations occurred in a range of clinical conditions and his intention was to differentiate the Charles Bonnet cases from visual hallucinations associated with parietal lesions, peduncular lesions and chronic hallucinatory psychoses. He also commented that the ocular lesions that one generally finds among these hallucinating old men are not the cause of their hallucinations:

“Contrairement à la théorie soutenue par les oculistes les lésions oculaires qu’on trouve le plus souvent chez ces vieillards hallucinés ne sont pas la cause de ces hallucinations”.

“Contrary to the theory supported by oculists, the ocular lesions that one generally finds among these hallucinated old men are not the cause of these hallucinations”.

In his later 1967 review de Morsier, insisted that eye disease be excluded from the definition since it was not the most important aetiologic factor; and emphasised instead the role of ageing on the cerebral visual pathways and the absence of a neuropsychiatric disorder:

“In 1938, j’ai proposé de désigner sous le nom de ‘syndrome de Charles Bonnet’ les hallucinations visuelles apparaissant chez les vieillards sans déficience mentale. Pour éviter toute confusion, il convient de conserver cette définition. Cette par erreur que quelques auteurs ont donné récemment ‘syndrome Charles Bonnet’ comme synonyme d’hallucinations chez des ophtalmobaphiles. Il n’existe pas de corrélation entre les hallucinations visuelles et les lésions des globes oculaires. Les hallucinations visuelles ne peuvent pas être expliquées par une privation d’afférences visuelles. Elles sont toujours causées par une altération du cerveau”.

In 1938, I tried to indicate under the name of “syndrome of Charles Bonnet” the visual hallucinations appearing among old men without mental deficiency. To avoid any confusion, it is convenient to preserve this definition. This is an error which some authors have recently applied to the “syndrome Charles Bonnet” as a synonym for hallucinations of the optic neuropathies. There does not exist a correlation between the visual hallucinations and the lesions of the ocular spheres. The visual hallucinations cannot be explained by a ‘dep- rivation’ of visual afferents. They are always caused by a disorder of the brain’.

de Morsier thus introduced an ambiguity. Charles Lullin and Bonnet himself both had eye disease, yet the CBS was not intended to describe this association. As a result, some authors follow de Morsier and reserve the Charles Bonnet eponym as a purely phenomenologi- cal description (complex hallucinations in the psychologi- cal normal) without specifying visual impairment and describe eye disease and old age as common clinical asso- ciations rather than diagnostic prerequisites. Other authors use the eponym to refer to those patients with complex visual hallucinations associated with eye disease.3,4 ffytche and Howard observed that at one extreme the term is used to describe:

1. All patients with complex visual hallucinations with preserved insight regardless of causal cerebral lesions, or visual impairment, while
2. for others the term is used to describe patients with complex visual hallucinations and eye disease. The sec- ond view is most generally accepted, with the proviso that often, there may be other operant factors. CBS appears to fall within the generalisation that patients with eye disease may experience the same distortions of visual perception as do those with cerebral lesions, and some normal subjects. One series reported screening 505 visually handicapped patients of whom 60 were found to meet proposed diagnostic criteria for CBS (generally, visu- al hallucinations without delusions or loss of insight).9,10

Patients have described seeing cartoon characters, flowers, hands rubbing each other, waterfalls and mountains, tigers, brilliantly coloured trees, street scenes, faces or life-size figures that they’ve never seen before often showing pleasant appearances. The underlying mechanism remains unclear but ffytche et al’s fMRI study of Bonnet’s syn- drome show that phasic increases in activity within specially visual cortex can underlie hallucinations of vivid colours and distorted faces.11 Indeed increases in visual cortical activity probably explain a variety of visual phe- nomena caused by different aetiologies that share a loss of inhibitory input.

References

Table 1: Inclusion criteria for the Charles Bonnet Syndrome

- At least one complex visual hallucination within the past 4 weeks;
- A period between the first and the last hallucination exceeding 4 weeks;
- Full or partial retention of insight into the unreal nature of the hallucinations;
- Absence of hallucinations in other sensory modalities;
- Absence of delusions.

Figure 1: Charles Bonnet

Figure 2: Title page: Essai Analytique Sur Des Faculte’s De L’ame, 1760.