European Journal of Ophthalmology / Vol. 19 no. 3, 2009 / pp. 494-495



# Acute reversible Charles Bonnet syndrome precipitated by sudden severe anemia

*PIERRE-FRANÇOIS KAESER, FRANÇOIS-XAVIER BORRUAT*

Hôpital Ophtalmique Jules Gonin, University of Lausanne, Lausanne - Switzerland

PURPOSE. *To report the sudden onset of reversible Charles Bonnet syndrome precipitated by acute severe anemia.*
METHODS. *The charts of three patients (Usher syndrome, bilateral macular degeneration, and bilateral retinal vein occlusion) with acute Charles Bonnet syndrome in the setting of severe anemia were reviewed.*
RESULTS. *Anemia resulted from bladder surgery, recto-colitis, and severe urinary tract infection. Hemoglobin ranged from 78 to 86 g/L. Decreased visual acuity and formed visual hallucinations (giants, flowers, animals) were present in all three patients. Rapid reversal of Charles Bonnet syndrome and visual acuity improvement followed blood transfusion.*
CONCLUSIONS. *Acute severe anemia can precipitate Charles Bonnet syndrome, which may be reversible by blood transfusion. (Eur J Ophthalmol 2009; 19:494-5)*

KEY WORDS. *Anemia, Charles Bonnet syndrome, Hallucinations*

*Accepted: October 23, 2008*

## INTRODUCTION

Charles Bonnet syndrome (CBS) is usually characterized by complex vivid visual hallucinations occurring spontaneously in people with intact cerebral function but with visual impairment. The insight into the unreality of the perceptions is always preserved in patients with CBS. CBS is estimated to occur in 11% to 15% of patients with visual impairment (1-4).
We report on three patients with Usher syndrome, bilateral macular degeneration, and bilateral retinal vein occlusion, in whom acute CBS was precipitated by sudden severe anemia. Further, the symptoms rapidly reversed following blood transfusion.

## METHODS

We reviewed the clinical charts of three patients with acute Charles Bonnet syndrome in the setting of severe anemia. This study was performed in accordance with the tenets of the Declaration of Helsinki.

## RESULTS

### *Case 1*

A 68-year-old man with Usher syndrome (retinitis pigmentosa and deafness) complained of visual loss and noticed the sudden onset of formed visual hallucinations 3 days after bladder surgery under general anesthesia. He described “people in front of a house surrounded by trees,” or sometimes, a “giant walking through the scene”. Vision was limited to hand motion (HM) bilaterally, whereas before surgery vision was counting fingers (CF) bilaterally. Severe anemia was present (hemoglobin 86 g/L). Blood transfusion resulted in rapid disappearance of hallucinations over 4–5 days and vision improved to CF bilaterally.

### *Case 2*

A 91-year-old woman with bilateral macular degeneration complained of a new decrease in vision accompanied by complex visual hallucinations 2 days after massive rectal bleeding due to rectocolitis. She saw a “field with a lot of

1120-6721/494-02$25.00/0

*Presented at the American Academy of Ophthalmology annual meeting, November 2006, Las Vegas, NV, USA.*

flowers, with the sun shining." The scene was "covered by intermittence by a lattice pattern." Visual acuity was 0.1 in the right eye, and CF in the left eye. Hemoglobin level was 80 g/L. Blood transfusions resulted in progressive decrease of visual hallucinations over the following week. Two weeks after the onset of visual hallucinations, visual hallucinations had disappeared, and visual acuity had improved to 0.5 in the right eye and 0.05 in the left eye.

### *Case 3*

A 90-year-old woman with bilateral cataract, macular degeneration, previous right branch retinal vein occlusion, and previous left central retinal vein occlusion complained of a drastic loss of vision and noticed the presence of formed visual hallucinations upon awakening from coma secondary to severe urinary tract infection. She mentioned seeing "animals in her room." Visual acuity was reduced to HM bilaterally. Hemoglobin level was 78 g/L. Following blood transfusion, visual hallucinations disappeared over 10 days. Visual acuity recovered to pre-coma values, 0.4 in the right eye, and CF in the left eye.

All three patients were always well oriented, and aware of the unreality of the perceptions. Bedside visual field examination was not suggestive of hemianopic defect. Neurologic examination was unremarkable. No cerebral imaging was obtained.

## DISCUSSION

Charles Bonnet syndrome is multifactorial, but is usually associated with bilateral visual impairment, mainly age-related macular degeneration, cataract, and diabetic retinopathy (3). The appearance of CBS depends more on the degree and speed of visual impairment rather than the underlying pathology (2). Restoration of vision (e.g. cataract surgery, vitrectomy) often results in cessation or improvement of the hallucinations (6, 7).

The mechanisms of CBS are unclear. Most theories postulate that loss of visual input alters the inhibition of self-generated cerebral activity. Anomalies of cerebral perfusion might also play a role in the generation of complex visual hallucinations (4).

The association with an external event other than visual acuity loss is not usually reported in CBS.

Our patients presented hallucinations associated with a new decrease of visual acuity in the setting of a severe and sudden anemia. Visual disturbances resolved after blood transfusion in all three cases. Two mechanisms in which anemia might be implicated in the pathogenesis of the cases are possible. First, further dysfunction of an already compromised macula, causing sudden loss of vision, might have increased cortical visual de-afferentation. Second, a transient cerebral ischemia might have resulted in further visual impairment. Neither neurologic dysfunction nor alteration of visual field suggestive of retrochiasmal dysfunction were found at bedside examination. Hence, a significant cerebral ischemia was unlikely.

The three cases we report here illustrate the importance of detecting anemia in the setting of acute visual hallucinations. Prompt correction of anemia is mandatory in order to prevent persistence of hallucinations and possible permanent visual failure.

*The authors have no proprietary or commercial interest in any materials discussed in this article. The authors have no financial support.*

Reprint requests to:
Francois-Xavier Borruat, MD
Hôpital Ophtalmique Jules Gonin
Avenue de France 15
CH-1004 Lausanne, Switzerland
francois.borruat@fa2.ch

## REFERENCES

1. Olbrich HM, Engelmeier MP, Pauleikhoff D, Waubke T. Visual hallucinations in ophthalmology. Graefes Arch Clin Exp Ophthalmol 1987; 225: 217-20.
2. Teunisse RJ, Cruysberg JR, Hoefnagels WH, Verbeek AL, Zitman FG. Visual hallucinations in psychologically normal people: Charles Bonnet's syndrome. Lancet 1996; 347: 794-7.
3. Teunisse RJ, Cruysberg JR, Verbeek A, Zitman FG. The Charles Bonnet syndrome: a large prospective study in The Netherlands. Br J Psychiatry 1995; 166: 254-7.
4. Menon GJ, Rahman I, Menon SJ, Dutton GN. Complex visual hallucinations in the visually impaired: the Charles Bonnet Syndrome. Surv Ophthalmol 2003; 48: 58-72.
5. Manford M, Andermann F. Complex visual hallucinations: clinical and neurobiological insights. Brain 1998; 121: 1819-40.
6. Rabins PV. The genesis of phantom (deenervation) hallucinations: an hypothesis. Int J Geriatr Psychiatry 1994; 9: 775-7.
7. Cogan DG. Visual hallucinations as release phenomena. Albrecht Von Graefes Arch Klin Exp Ophthalmol 1973; 188: 139-50.