ARCH SOC ESP OFTALMOL. 2017;xxx(xx):xxx–xxx

ARCHIVOS DE LA SOCIEDAD ESPAÑOLA DE OFTALMOLOGÍA

www.elsevier.es/oftalmologia

Short communication

# Charles Bonnet syndrome in a child with congenital glaucoma☆

*E. Santos-Bueso*, M. Serrador-García, F. Sáenz-Francés, C. Méndez-Hernández, L. Morales-Fernández, J.M. Martínez-de-la-Casa, J. García-Feijoo, J. Porta-Etessam*

*Unidad de Neurooftalmología, Servicio de Oftalmología, Instituto de Investigación Sanitaria, Hospital Clínico San Carlos (IdISSC), Madrid, Spain*

ARTICLE INFO

*Article history:*
Received 14 September 2016
Accepted 9 November 2016
Available online xxx

*Keywords:*
Charles Bonnet
Syndrome
Hallucinations
Child

ABSTRACT

*Case report:* The case is presented of a 12 year-old boy with congenital glaucoma and low visual acuity diagnosed with Charles Bonnet syndrome. This consisted of the acute onset of complex, repetitive, persistent, and with visual hallucinations (people, brooms and coffeemakers) of self-limited evolution without treatment. The patient was diagnosed with congenital glaucoma at 3 years of age, and subjected to a trabeculectomy in right eye, and trabeculectomy and keratoplasty in his left eye.

*Discussion:* Charles Bonnet syndrome symptoms have been described in adults, but their presence in children is poorly reflected in literature, with unknown characteristics and prevalence.



## Síndrome de Charles Bonnet en un niño con glaucoma congénito

*Palabras clave:*
Charles Bonnet
Síndrome
Alucinaciones
Niño

RESUMEN

*Caso clínico:* Presentamos el caso de un niño de 12 años de edad con glaucoma congénito y baja agudeza visual, diagnosticado de síndrome de Charles Bonnet, consistente en alucinaciones visuales complejas (personas, escobas y cafeteras), repetitivas, persistentes y de aparición brusca, con evolución autolimitada sin tratamiento. El paciente había sido diagnosticado de glaucoma congénito a los 3 años de edad e intervenido mediante trabeculectomía en ojo derecho y trabeculectomía y trasplante de córnea en ojo izquierdo.

☆ Please cite this article as: Santos-Bueso E, Serrador-García M, Sáenz-Francés F, Méndez-Hernández C, Morales-Fernández L, Martínez-de-la-Casa JM, et al. Síndrome de Charles Bonnet en un niño con glaucoma congénito. Arch Soc Esp Oftalmol. 2017. http://dx.doi.org/10.1016/j.oftal.2016.11.008

* *Corresponding author.*
E-mail address: esbueso@hotmail.com (E. Santos-Bueso).

*Discusión:* El síndrome de Charles Bonnet ha sido descrito en adultos, afecta a más del 15% de los pacientes con baja visión, pero su presencia en la edad pediátrica es escasamente reflejada en la literatura, y se desconocen sus características y su prevalencia.



## Introduction

The presence of visual hallucinations in patients is on the increase in our environment. It is estimated that approximately 15% of patients with low vision experience an episode of the Charles Bonnet syndrome[1] (CBS). Hallucinations experienced by patients with poor vision and preserved cognitive condition, constituting CBS, are very infrequent in the pediatric age and scarcely documented.[2,3]

The case of a 12-year-old child with congenital glaucoma is presented, who exhibited a self-limited CBS during 6 months which did not require medical treatment.

## Clinical case

Male, 12, who referred predominantly nocturnal complex visual hallucinations consisting in visions of unknown people who walked around him without interacting, as well as vision of objects (a coffee maker and a broom) in black and white, which appeared with a frequency of approximately 2–3 times a week. The patient interpreted said visions as unreal and was aware of his sensory deficit. No relevant familial history or known allergy was referred. The patient had been diagnosed with congenital glaucoma at age 3, had undergone trabeculectomy in both eyes (BE) and subsequently penetrating keratoplasty in right eye (RE) and Ahmed valve implant in left eye (LE). Examination produced finger counting at 1 m in RE and 20/200 in LE that did not improve with stenopeid hole in BE and intraocular pressure of 14 mmHg in BE. OCTOPUS campimetry was performed (Fig. 1) which showed concentric defect in RE and nasal step in LE. The patient was examined at the Neuro-Ophthalmology Unit and diagnosed with CBS. It was decided not to administer medical treatment as the syndrome did not produce symptoms such as anxiety or fear as well as because the patient was self-critical of his hallucinations. Six months later, said visual hallucinations disappeared spontaneously.

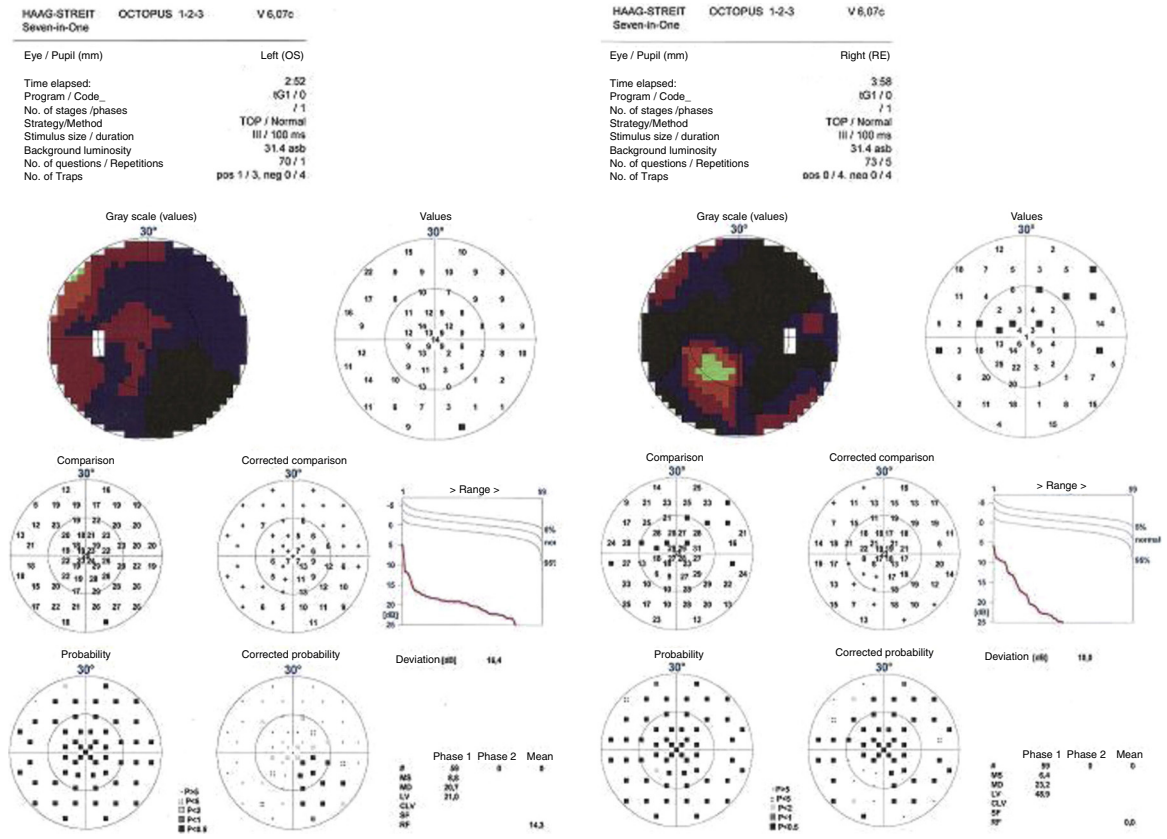


**Fig. 1 – Patient campimetry with concentric reduction in right eye and nasal step in left eye.**

## Discussion

The estimated prevalence of visual hallucinations in patients with poor vision ranges between 11 and 15%,[1] However, studies reported lower prevalence, possibly due to the lack of knowledge about CBS and the fear of patients to be classified as psychiatric cases.[1,4–6] Most cases have been described in elderly patients between 75 and 84 years of age, with slight predominance of females, who exhibited age-related visual diseases such as macular degeneration, glaucoma or cataracts. However, CBS has also been described in patients with good vision, possibly related to deafferentiation of damaged fibers despite preserving central vision.[1,7]

A number of theories have been proposed to explain the origin of hallucinations. When visual acuity is diminished, the visual cortex stimulation from the retina is also diminished without complete disappearance of neuronal activity, as occurs in amaurosis. This gives rise to what is known as the deafferentiation process that produces hyper-excitability of a specific cortical area in an offsetting effort by the deafferentiated brain area that produces anatomical as well as biochemical offsetting changes.[1,4–7]

CBS in pediatric patients is scarcely reported in the literature. Only 2 studies were found,[2,3] excluding the possibility of establishing a prevalence rate. Mewasingh et al.[2] describe the case of a 9-year-old patient with severe visual impairment secondary to craniopharyngioma, who exhibited visual hallucinations of animals (zoopsia). Said authors also included references of 3 published cases secondary to cone dystrophy and optic nerve glioma.[2] Schwartz et al.[3] described the cases of 2 children, 6 and 8 years old, with visual impairment secondary to cone dystrophy who exhibited hallucinations consisting in buildings, human beings and geometrical figures (patient 1) and vision of colored spheres, human beings and monsters (patient 2).

In what concerns CBS treatment, there is a large range of drugs that act at different levels of the central nervous system which must be managed by multidisciplinary departments comprising Neurology and Ophthalmology.[8] Due to the scarcity of statistics there are no guidelines or protocols available. In the case presented herein, hallucinations were self-limited and disappeared 6 months later without requiring treatment because the patient was critical of them and his daily life was not disturbed. Mewasingh et al.[2] describe that the patient was treated with carbamazepine due to the epileptic crises associated to the tumor which diminished the frequency of hallucinations.

Awareness about CBS, specifically in pediatric patients, when its prevalence is significantly lower, is important to develop an adequate differential diagnostic in order to optimize diagnostic test and treatments, considering the particular vulnerability of said age group. Both children and their families would benefit from adequate information about the Charles Bonnet Syndrome.[1,4–8]

## Conflict of interests

No conflict of interests was declared by the authors.

## REFERENCES

1. Santos-Bueso E, Sáenz-Francés F, Serrador-García M, Porta-Etessam J, Martínez-de-la-Casa JM, García-Feijoo J, et al. Prevalence and clinical characteristics of Charles Bonnet syndrome in Madrid, Spain. Eur J Ophthalmol. 2014;24:960–3.
2. Mewasingh LD, Kornreich C, Christiaens F, Christophe C, Dan B. Pediatric phantom vision (Charles Bonnet) syndrome. Pediatr Neurol. 2002;26:143–5.
3. Schwartz TL, Vahgei L. Charles Bonnet syndrome in children. J AAPOS. 1998;2:310–3.
4. Menon GJ, Rahman I, Menon SJ, Dutton GN. Complex visual hallucinations in the visual impaired: Charles Bonnet syndrome. Surv Ophthalmol. 2003;48:58–72.
5. Burke W. The neural basis of Charles Bonnet hallucinations: a hypothesis. J Neurol Neurosurg Psychiatry. 2002;73:535–41.
6. Teunisse RJ, Cruysberg JR, Hoefnagels WH, Verbeek A, Zitman G. Visual hallucinations in psychologically normal people: Charles Bonnet's syndrome. Lancet. 1996;347:794–7.
7. Madill SA, Ffytche DH. Charles Bonnet syndrome in patients with glaucoma and good acuity. Br J Ophthalmol. 2005;89:785–6.
8. Santos-Bueso E, Serrador-García M, García-Sánchez J. Treatment of Charles Bonnet syndrome. Arch Soc Esp Oftalmol. 2013;88:244–5.