Lesson of the week

Fear of the dark in children: is stationary night blindness the cause?

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Fear of the dark is a common complaint of pre-teenage children.[1 2] It should not be confused with night terrors or panics, in which a child becomes acutely agitated and terror-struck at night, appearing to be awake while in fact asleep and unable to be woken.[3 4] In contrast, fear of the dark can be experienced by the conscious child in dimly lit or dark conditions. When such fear is excessive it is often attributed to attention seeking behaviour or assumed to be an irrational fear that will abate with time.

Most people can see a little in very dim lighting conditions after a short period of adaptation. However, a child with no visual problem obvious to the parents and who can see normally in well lit conditions can present as being unable to see at all in the dark even after a period for adaptation. In a child who cannot yet talk this may simply appear as fear of the dark. We describe two patients with congenital stationary night blindness, a diagnosis which may be missed without appropriate history taking from the parents, particularly if there is no family history of visual problems.

Case 1

A 3 year old girl had been frightened of the dark from an early age. She had complained to her parents of not being able to see when the bedroom lights were turned off, prompting the natural response, "Wait until your eyes adjust." She would persist with her complaints of not seeing and her parents would switch her light back on again. She had had profound problems getting to sleep and would not leave her bedroom in the dark, although she would move from her dark bedroom into the lit hallway. She also refused to enter her parents' darkened bedroom.

Her younger sister was born with a history of being visually unresponsive from birth and was referred at the age of 3 months for ophthalmological investigation. She was found to have reduced vision, nystagmus, and long sightedness but a normal ocular examination. Her vision improved considerably after glasses were prescribed, but electrophysiological testing suggested abnormal retinal function. This finding prompted the parents to seek investigation of their older daughter.

The older child's vision was 6/12 in each eye with no nystagmus and normal ocular examination. Her pupils responded by dilating to a bright light (paradoxical pupil response). Electrophysiology showed a "negative" electroretinogram, consistent with a diagnosis of congenital stationary night blindness.

An intensity adjustable night light was suggested and within days she was more confident and sleeping through the night. It was also suggested that she could carry a torch when going out at night and that her parents should draw the curtains before dusk to avoid her fear of the darkness beyond the windows.

Case 2

A girl of 2 was referred with a history of frequently tripping over and bumping into objects as well as problems seeing in the dark. Her parents reported that when she woke at night she would cry but would not go to her parents' bedroom and was very frightened. She also needed to have curtains drawn at night because of her fear of the darkness beyond the windows. Examination revealed her to have normal visual acuity and slight long sightedness. She had pendular nystagmus, but pupillary and fundus examination showed nothing abnormal. There was a family history of nystagmus and impaired night vision. Electrophysiology revealed a "negative" electroretinogram consistent with a diagnosis of congenital stationary night blindness. Now that she is in control of her lighting by having a bright hall light shining into her bedroom at night and by carrying a torch outside at night, her fear of the dark is no longer evident. She does, however, still have some problems with shadows on the wall, which appear to her to be holes. Her parents have got her to feel the shadows with her hands moving from the light areas to the dark to appreciate the continuity of the solid wall while explaining to her the nature of shadows cast by objects in front of a light.

Discussion

There are many causes of nyctalopia (night blindness), which can be divided into stationary forms such as congenital stationary night blindness, fundus albipunctatus, fleck retina of Kandori, and Oguchi's disease[5 6] and the more numerous and severe progressive forms such as retinitis pigmentosa, Refsum's disease, choroideremia, vitamin A deficiency, and retinopathy associated with cancer.[6–8]

Congenital stationary night blindness is rare and its prevalence has not been well established, although it is about three times more common in boys than in girls, probably reflecting the presence of an X linked mode of inheritance in addition to the autosomal dominant and recessive forms.[9]

Congenital stationary night blindness is an inherited, non-progressive disorder which affects principally rod photoreceptor function in the retina, resulting in impaired night vision, although there is commonly a mild impairment in cone function, which may affect daytime central visual acuity.[10–13] The defective pathway appears to result from a signal transmission defect from the photoreceptors. It principally manifests itself as an increase of the dark adapta-

Fear of the dark in children may have a pathological basis

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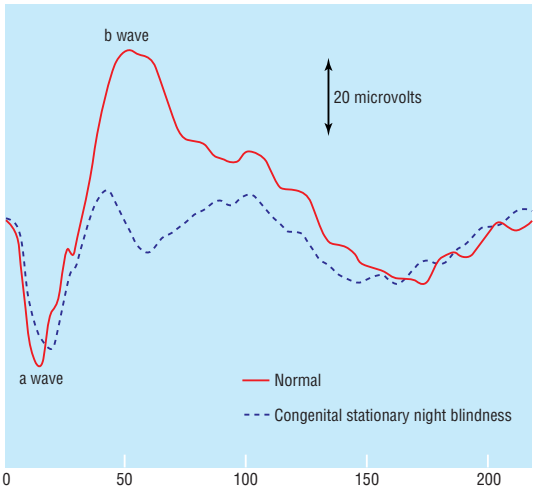
BMJ 2003;326:211–2



Mixed rod-cone electroretinograms for normal individual and patient with congenital stationary night blindness. The *a* wave is generated by photoreceptors; the *b* wave is the result of subsequent depolarisation of cells in the inner nuclear layer of the retina. In one form of congenital stationary night blindness the *a* wave is preserved, but the *b* wave is attenuated, giving rise to the negative electroretinogram shown here

tion threshold, nystagmus, and often moderate to high short sightedness, although the refractive state may be long sighted or normal. Squint, pale or tilted optic discs, and paradoxical pupil responses have all been reported in association with the condition.[6] There is usually no visual deficit in good lighting conditions but mild central visual loss is common in some inherited forms. Fundus examination is usually normal, distinguishing it from the progressive conditions causing nyctalopia. The diagnosis can be established by electroretinography.

There are several well known congenital stationary night blindness pedigrees worldwide with varied features,[14 15] but despite the phenotypic variability between patients three characteristics appear to be consistent: night vision impairment; raised or absent rod thresholds; and absent or diminished electroretinogram *b* waves, creating a "negative" waveform (figure).[12]

The key to diagnosing congenital stationary night blindness is to listen carefully to the history and to ask in particular about a family history of visual problems. Our patients both showed the same features: fear of the dark, problems with mobility at night, and insisting that curtains were drawn much earlier than others would choose. One child had fear of shadows. A detailed history should be followed by a complete ocular examination and electroretinography. Once congenital stationary night blindness has been diagnosed there are several strategies that parents can adopt to help their children cope with and be in control of their environment. There are also manuals available to parents with strategies to help reduce a child's fear of the dark.[2] In addition, genetic counselling may be helpful for parents with affected offspring. Night blindness in children can bring about profound fear of the dark. Recognition of the diagnosis and the simple expedient of giving the child control of lighting conditions can transform family life.

Contributors: SSS drafted the manuscript. RH provided the electrophysiology and helped to draft the manuscript. GND made the diagnoses, had the idea for the paper, and commented on the draft manuscript.

Funding: None.

Competing interests: None declared.

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(Accepted 21 March 2002)

Corrections and clarifications

Visible signs of illness from the 14th to the 20th century: systematic review of portraits

In this article by C Als and colleagues in the Christmas issue of the *BMJ* (21-28 December, pp 1499-501), we added the wrong affiliation for one of the authors. Y Stüssi is a medical assistant at the Burgerbibliothek Bern, in Berne, Switzerland, not the director, as stated.

Photofinish

And better late than never—here's a correction to the Photofinish section of the Christmas issue of 2001. Three authors' names should have been published along with P Shah's for the picture submitted about the "smiling face" defect (*BMJ* 2001;323:1498). The missing authors—all from the Birmingham and Midland Eye Centre, City Hospital NHS Trust, Birmingham B18 7QU—are B Mushtaq, senior house officer in ophthalmology; V Kumar, specialist registrar in ophthalmology; and U S Ramnathan, senior fellow in ophthalmology.

Endpiece

Mark Twain on evidence based practice

It ain't what people don't know that hurts them it's what they know that ain't so.

Mark Twain