Fear of the dark is a common complaint of pre-teenage children. 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. 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 albinopathy, fleck retina of Kandori, and Oguchi’s disease and the more numerous and severe progressive forms such as retinitis pigmentosa, Refsum’s disease, choroideremia, vitamin A deficiency, and retinopathy associated with cancer.

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.

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. 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-
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.

Corrected 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.

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**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