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Transient, Unexplained, and Psychogenic Visual Loss in Children

Introduction

Some children have visual disturbances that occur in the absence of, or are out of proportion to, their objective ophthalmological findings. These symptoms reflect a wide range of processes that may be benign or may be a sign of neurological, systemic, or psychiatric disease. This chapter deals with the neuro-ophthalmologic detection of organic and psychogenic disorders that may manifest as transient or unexplained visual loss in childhood.

Transient Visual Disturbances in Children

Most of what is known about transient visual disturbances has come from correlating pathology and pathophysiology from the richly detailed descriptions given by some adults. Formed and unformed visual hallucinations have occurred in artists who have been able to paint or draw what they have seen. Physicians and scientists have experienced and have described many visual disturbances, such as amaurosis fugax and the scintillating scotoma of migraine. Many of the same episodic visual disturbances occur in childhood, but several formidable problems confront the physician trying to reach the correct diagnosis. The descriptions of episodic visual disturbances and hallucinations in children are less complex in detail than those of the adult population, because children have a limited vocabulary and a limited experiential basis of sensory phenomenon to draw upon.

Children share with adults a difficulty in distinguishing homonymous from monocular defects and may insist that a homonymous defect affects only one eye. Children are also less likely than adults to draw a distinction between a positive and a negative visual disturbance. They may simply maintain that something is blocking their vision and may be unable to describe it further. If pressured by the examiner to be more descriptive, a child or even a teenager may attempt to give the examiner what they think is being asked for, even if it does not accurately depict their symptoms. Children with refractive errors or other organic visual problems may describe visual symptoms only in terms of their effect upon a specific activity (such as difficulty reading the blackboard at school or reading textbooks), making it difficult to determine from the history whether the disturbance is indeed episodic.

The commonest cause of episodic visual loss disturbances in childhood is migraine. The visual disturbance of migraine is characterized by episodic visual hallucinations and visual loss, as well as other neurological disturbances, with headache being the most common. However, the characteristic hemicranial throbbing headache is often absent in the pediatric age group, and the diagnosis is based upon a compilation of circumstantial evidence. A personal profile of the child should be explored with specific attention to eliciting a history of extreme fussiness or colic as a baby, night terrors, recurrent abdominal pains, or motion sickness.[40] A family history of migraine must be sought since family members with migraine may never have been diagnosed or may have been mis-

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diagnosed as having tension or sinus headaches. The diagnosis of migraine for the child's visual disturbances and the parent's headaches can often be established in the same interview.

Careful questioning may determine that the child is describing a visual hallucination rather than a visual obscuration. As in adults, visual hallucinations in children may be formed or unformed and may be simple or complex. Unformed hallucinations typically consist of lights, heat wave sensations, or simple geometric patterns that may be spatially stable or move. Formed hallucinations consist of recognizable objects or people. These may be simple, such as visualizing a single animal or an object, such as a table or chair, or they may demonstrate varying degrees of complexity involving the purposeful movement of several people in a scene with appropriate colored backgrounds and facial expressions. If the attacks are repetitive, the hallucination may be stereotyped, or a new scene or object may be visualized with each recurrence.

Visual hallucinations are generally divided into *irritative* and *release* hallucinations.[74] Irritative hallucinations are usually caused by epileptic discharges that occur as part of a seizure.[122,174] Irritative hallucinations emanating from the temporal lobes tend to be complex and stereotyped, while those arising in the occipital lobes tend to be simple and unformed. Other aspects of the seizure disorder are often more prominent, including changes in consciousness and sensory or motor abnormalities due to the spread of the epileptic activity; however, isolated and localized occipital or temporal lobe seizures may produce visual hallucinations as their only manifestation.

Release hallucinations often occur in patients with decreased vision or visual field defects. They may also occur in the setting of monocular or binocular visual loss or homonymous hemianopia and may manifest in patients with relatively mild visual loss.[36] These hallucinations range from unformed phosphenes to formed hallucinations with complex patterns. Release hallucinations presumably occur when normal visual impulses are removed, releasing indigenous cerebral activity within the visual system.[36] They tend to be continuous and can last from minutes to days, in contradistinction to irritative hallucinations that last for seconds to a few minutes.[36] Release hallucinations are neither associated with electroencephalographic abnormalities nor altered by anticonvulsant therapy.

The failure to clearly distinguish the irritative from the release type of hallucination has led to considerable confusion regarding their localizing value. The concept that hallucinations of occipital origin comprise unformed phosphenes applies only to the irritative variety. Unlike irritative hallucinations, which vary in character depending upon their site of origin, release hallucinations have no localizing value and can follow injury to the visual system anywhere from the eye to the occipital cortex.[36,130,237] For example, formed release hallucinations occasionally occur in adults with dense cataracts or macular degeneration.[139] Children can experience "phantom vision" following enucleation of one or both eyes.[37] Patients with visual loss frequently acknowledge experiencing both formed and unformed visual hallucinations when specifically asked.

Migrainous Phenomena

Migraine is not just a headache.[31] It is a disorder that can cause transient sensory, autonomic, motor, visual, and cognitive impairment. Although headache is a prominent feature of migraine, it is not invariably present.[31] Many migraine attacks begin slowly and evolve through sequential stages of neurological dysfunction. Selby[202] described migraine as a "drama in three acts," comprising prodrome (frequently not recognized) and aura, a headache phase, and a post-headache phase. The aura may occur prior to, concurrently, or even after the onset of the headache. It cannot be overemphasized that the diagnosis of pediatric migraine is established on the basis of the personal profile, attack profile, and family history, as well as the absence of physical findings. In the child with transient visual disturbances or unexplained headache, the personal history often provides an important clue to the diagnosis of migraine.[40] Pediatric migraineurs often have a history of migraine equivalents, including colic, recurrent abdominal pain, cyclic vomiting, pavor nocturnus (night terrors), and paroxysmal torticollis within the first few years of life.[15,31] Even between attacks, migraineurs often describe