Developmental Outcomes of Shaken Baby Syndrome: A Case Report
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ABSTRACT

Background: Shaken baby syndrome is a form of non-accidental trauma caused from violently shaking a child. The classic triad of findings includes retinal hemorrhages, subdural hemorrhages, and encephalopathy. In addition to these symptoms, many long-term outcomes are related to physical disabilities, cognitive impairments, behavior disorders, and visual processing disorders.

Case Summary: An 11-year-old African-American male presented for a comprehensive eye examination with the complaint of an eye turn. The patient suffered from shaken baby syndrome as an infant, which resulted in swelling of his right optic nerve. As a result, the patient had right optic nerve atrophy, a constant right exotropia, myopic anisometropia, and decreased best-corrected visual acuity. A new glasses prescription was written, and the parents were informed of the importance of regular comprehensive eye exams and possibilities for visual aids/therapy to help with his activities of daily living.

Discussion: Developmental outcomes related to shaken baby syndrome can include many visual and behavioral disorders. Additionally, patients with a history of shaken baby syndrome typically show problems with visual memory, cognitive impairment, and behavioral disorders.

Keywords: Shaken baby syndrome, non-accidental head trauma, abusive head trauma, visual perceptual deficit

Introduction
Shaken baby syndrome is a form of non-accidental trauma caused by violently shaking a child. In the United States, the incidence of trauma in infants is roughly 20 cases per 1000 children per year. The leading cause of death and disability in this group is related to brain injury. Typically, boys are more often affected than girls, and most injury occurs before one year of age, with the peak incidence being at 10-16 weeks of age.

In shaken baby syndrome, the child is violently shaken, causing a classic triad of findings: retinal hemorrhages, subdural hemorrhages, and encephalopathy (Table 1). Additional findings include lethargy, irritability, difficulty breathing, inability to lift the head, and poor eye tracking, among others (Table 2). Shaken baby syndrome is thought to occur from the acceleration, deceleration, and rotational forces that occur during violent shaking with or without impact. There is some debate as to whether the signs and symptoms of shaken baby syndrome can actually occur related to violent shaking. In 2009, the American Academy of Pediatrics adopted the term “non-accidental head injury” to describe what is commonly known as shaken baby syndrome.

Ocular findings are important to the diagnosis of shaken baby syndrome because these findings do not typically occur with other forms of brain injury in the infant and toddler populations. Retinal hemorrhages can occur at all layers of the retina and are present in approximately 80% of cases. In addition, the patient may present with preretinal or vitreous hemorrhages, papilledema, and macular retinoschisis. If not monitored or treated properly, these complications may lead to visual impairment.

Eye care professionals may be the first individuals to diagnose abuse, and it is important that any cases of suspected child abuse are reported. Ocular findings that might indicate child abuse include retinal hemorrhages, dislocated intraocular lens(es), bruising around the eyes, and conjunctivitis caused by a sexually transmitted disease. Additionally, an eye care professional may notice bruising in areas where children are not commonly injured, multiple bruises in different stages of healing, burn or bite marks, and unexplained fractures. Practitioners should be aware of the reporting requirements.
within their area of practice in addition to support services for the children and other family members.

Long-term follow-up of children with a history of shaken baby syndrome can be challenging because these children may be in foster care or may move frequently. These children may not see the same physician regularly, and their medical and ocular history may not be known by their foster or adoptive parents. Because of these challenges, long-term outcomes from abusive head trauma may be difficult to identify and to report in the literature. Some of the long-term consequences may include learning, physical, and visual disabilities, as well as behavior disorders and cognitive impairment.

The case reported in this article discusses the outcome of one child who suffered abusive head trauma as an infant. It details the various ocular and developmental outcomes that are possible with this patient population, as well as the treatment recommendations for the visual and developmental deficits.

Case Summary

An 11-year-old African-American male presented for an eye examination with his adoptive mother and father. The patient’s chief complaint was an eye turn, which had been present since birth. The parents felt that his glasses helped with the eye turn. History revealed that the patient suffered from shaken baby syndrome when he was an infant. This non-accidental trauma resulted in a hematoma, causing decreased vision in his right eye. He also had a water bottle burn scar on his stomach and a history of a broken left leg when he was 2 months old. The patient had been with his adoptive parents since he was 6 months old, and they reported that he was born full-term via a C-section delivery.

He presented with aided acuities of OD 20/400, OS 20/30 at distance and OD 20/150, OS 20/60 at near through his habitual prescription of OD -8.50-2.25x010, OS -1.25-4.25x170. Pupils, extraocular motility, and confrontation visual fields were all normal. Cover test revealed an 8° constant right exotropia which was equal at distance and near. The subjective refraction yielded a prescription of OD -7.50-3.50x005, OS +0.25-4.50x170 with visual acuities in the distance of OD 20/400, OS 20/20-. The new prescription did not change the cover test findings.

Anterior segment assessment was unremarkable for both eyes. The patient’s intraocular pressures were 19mmHg OD, 18mmHg OS. Posterior segment assessment of the right eye revealed a vertically elongated optic nerve head with optic atrophy and a C/D ratio of 0.2H/0.3V (Figure 1). No foveal light reflex was present in the right eye. The posterior segment assessment of the left eye was unremarkable with a C/D ratio of 0.4H/0.4V and a present foveal light reflex.

Discussion

The long-term effects of shaken baby syndrome are numerous and variable. Typically, children who suffer from a traumatic brain injury such as shaken baby syndrome at a younger age tend to have more difficulties later in life. Long-term effects can occur in a variety of different areas. Children may show ocular and visual processing complications, physical disabilities, cognitive impairment, and behavioral disturbances.

Ocular Complications

The effects of non-accidental head injury can last after the physical signs of the injury have disappeared. It is estimated that approximately half of children who have suffered from non-accidental head injury have lasting visual abnormalities, including cortical blindness, decreased visual acuity, and visual field deficits.

In the case presented, the patient had reduced visual acuity of his right eye. Without having early health records, one can only theorize why this occurred. When a visual disturbance such as a congenital cataract occurs in an infant, the eye elongates as a result of the visual deprivation. As the eye elongates, the refractive error moves further away from emmetropia. With this case, the child had swelling of the right optic nerve that eventually resulted in optic atrophy. This likely caused an early visual deprivation, which then caused the axial length of the right eye to increase. The reduced visual acuity in the right eye could be related to anisometric refractive amblyopia in addition to the right eye optic atrophy. The subjective refraction was prescribed for this patient with the recommendation of a protective lens material. Amblyopia therapy could have been used to see if there was any potential for improving the acuity in the right eye. Because the acuity in the right eye was significantly decreased, traditional patching therapy would need to be done at home only so as not to limit the child’s ability to perform well in the classroom. With the additional complication of the optic atrophy, it is difficult to hypothesize how much visual acuity improvement could be expected with amblyopia therapy.

In addition to the poor acuity in the right eye, the patient had a small-angle constant right exotropia which was of equal

Figure 1. Picture of the right optic nerve showing atrophy and a vertically elongated appearance.
magnitude at distance at near. Sensory strabismus occurs when there is significant vision loss that inhibits sensory fusion.\textsuperscript{11} Without having access to early records, it is difficult to determine whether the constant right esotropia was present prior to the trauma. Based on the information available, one can postulate that the exotropia was a sensory strabismus to allow for suppression of the eye with the worse best-corrected visual acuity. When impairment leading to poor sensory fusion occurs prior to the age of 5, as it did in this case, the direction of the deviation has been found to be equally distributed between esotropia and exotropia.\textsuperscript{11} However, when impairment to sensory fusion occurs later in life, the direction of the deviation is more commonly outward, resulting in an exotropia. In cases such as this one, the underlying cause of the sensory strabismus limits the ability to regain sensory fusion through therapy. To help correct the appearance of the strabismus, prisms can be placed in the glasses for smaller deviations. For larger-angle deviations, extraocular muscle surgery is an option to help cosmetically align the eyes. Although the patient did exhibit a full range of motion of his extraocular muscles (EOMs), the development of the innervation to the EOMs may have been disrupted during the non-accidental trauma.

**Visual Processing Deficits**

Brain injury at a young age can lead to a variety of visual processing deficits, including problems with visual memory, visuospatial skills, and visual agnosia.\textsuperscript{9,12-14} Most of these deficits may not be noted until the child begins school, and even then, some of these deficits can exist for many years before causing significant problems.

No visual processing testing was performed on this child. The adoptive parents reported that the child was doing well in school, although he had a learning disability. The parents were not interested in further testing for a learning-related vision disorder. Based on previous studies, children sustaining a brain injury at an earlier age, as well as a more severe injury, are more likely to suffer from visual processing deficits.\textsuperscript{9,12-14} Based on this patient’s history of trauma within the first few months of life, he may have deficits in one or more areas of visual processing.

**Physical Disabilities**

When infants are admitted to the hospital for shaken baby syndrome, they are likely to have physical findings, including rib fractures and skeletal fractures.\textsuperscript{9} As these children grow and develop, these findings, along with the brain injury, have an effect on their physical development. Slightly more than half of post-shaken baby syndrome children show some type of gross motor difficulty, although functional mobility is normal in 60% of children.\textsuperscript{8}

In the case presented, the patient had a broken leg when he was 2 months old caused by non-accidental trauma, in addition to a water bottle burn on his stomach. The patient’s gross mobility was normal; he was able to walk into the examination room and sit comfortably in the examination chair. He did not appear to have an abnormal gait, and the parents did not report any physical limitations.

**Cognitive Impairment**

Infants and children who sustain a brain injury at a young age are more likely to show cognitive deficits than children who sustain a brain injury at an older age.\textsuperscript{13,14} The brain injury interferes with the acquisition of new skills, which can then lead to cognitive impairment predominantly in the areas of memory, attention, rapid motor performance, and academic skills development.\textsuperscript{14}

Infants and children who suffer from a brain injury tend to have a slower rate of learning compared to their non-brain injured classmates. This has an effect of poor word decoding when reading and a failure to use language to communicate effectively.\textsuperscript{14} Additionally, children who suffer from a brain injury at a younger age show a lower IQ and less recovery of IQ as the child ages.

For this patient, the adoptive parents noted that he had a learning disability that they attributed to the shaken baby syndrome. The child was able to read and was in regular education classrooms. The parents were not specific about the areas in which the patient suffered, but they stated that he had gone through educational testing, and they were not interested in further testing in this area.

**Behavioral Disturbances**

As with many other long-term consequences, behavioral disturbances may not be apparent until the child reaches school age or adolescence. Common abnormalities include problems with sleep, hyperactivity, impulsivity, temper tantrums, and rage reactions.\textsuperscript{9} Many of these children have problems socializing with their peers due to poor language skills, and this may lead to behavioral problems at school and in social situations.

No behavioral problems were noted by the adoptive parents, and the patient appeared to be well-behaved during the examination. He was able to follow directions with minimal reminders, and he was never agitated or outspoken during the 2-hour examination.

While this case report details some of the common long-term outcomes from abusive head trauma, it can be difficult to determine the potential of an individual patient. In some patients, the visual system may be fully functional, but the child may not be able to recognize and to communicate what they are seeing. Conversely, the child may have visual system deficits but a normal intellectual ability. This case report details the findings from one patient and can be used to guide professionals in what areas to evaluate further. However, it should not be taken as an exhaustive list of all possible complications or outcomes from non-accidental head trauma.
Conclusion

Shaken baby syndrome can have an effect beyond the infancy years. Children who have suffered brain injury at an early age are more likely to have cognitive, behavioral, and visual processing problems than their peers, and it is important for optometrists to be aware of these possible findings when examining children with a history of shaken baby syndrome or other brain injury at an early age. The patient presented in this case suffered from ocular and visual processing complications and a learning disability. He received optical correction and was offered further visual processing testing. At this time, the adoptive parents decline further intervention.

References


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