

INFORMATION STATEMENT

Abusive Head Trauma/Shaken Baby Syndrome June 2010

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Although most eye injuries in childhood are a result of accidents, a significant portion result from physical abuse by adults. Child abuse, including physical abuse, sexual abuse, neglect, and emotional abuse, is a pervasive problem in our society, with an estimated 1,256,600 million confirmed victims per year in the United States alone (http://www.acf.hhs.gov/programs/opre/abuse_neglect/natl_incid/index.html). Physically abusive behavior by a parent or other caregiver usually reflects temporary loss of control during a period of anger or stress rather than premeditated cruelty. Lack of knowledge of normal child development and the proper way to care for or discipline a child are also frequent contributing factors.

A reliable history is often difficult to obtain when child abuse has occurred. Suspicion should be aroused when repeated accounts of the circumstances of injury or histories obtained from different individuals are inconsistent or when the events described seem to conflict with the extent of injuries (e.g., bruises on multiple aspects of the head after a fall) or with the child's developmental level (e.g., a 2 month old rolling off a bed or a 9 month old climbing out of a high chair). Any physician who suspects that child abuse might have occurred is required by law in every US state and Canadian province to report the incident to a designated governmental agency. Once this obligation has been discharged, the ophthalmologist may be an important contributor to understanding the case but probably is best advised to leave the full investigation of the situation to appropriate specialists or other authorities.

The presenting sign of child abuse involves the eyes in approximately 5% of cases, and ocular manifestations are detected in the course of evaluating many others. Blunt trauma inflicted with fingers, fists, or implements such as belts or straps is the usual mechanism of nonaccidental injury to the ocular adnexa or anterior segment. Periorbital ecchymosis, subconjunctival hemorrhage, and hyphema should raise suspicion of recent abuse if the explanation provided is less than completely plausible, particularly in infants. Cataract or lens dislocation may be signs of injury or trauma inflicted in the past. A majority of rhegmatogenous retinal detachments that occur in childhood have a traumatic origin; abuse should be suspected when such a finding is encountered in a child without a history of injury or an apparent predisposing factor such as high myopia. Certain signs of trauma include ruptured globe, orbital fracture, vitreous base detachment and retinal commotio. Trauma should also be considered in the differential diagnosis, of unilateral optic atrophy, hyphema, and periocular ecchymosis, in the absence of another clear explanation.

Shaken Baby Syndrome

Shaken Baby Syndrome is a subset of abusive head trauma characterized by repetitive acceleration-deceleration forces with or without blunt head impact resulting in a unique complex of ocular, intracranial, and sometimes other injuries, usually in infants. Because the essential features were identified in the early 1970s, it has become widely recognized as one of the most serious manifestations of child abuse.

Abusive Head Trauma/Shaken Baby Syndrome

Victims of shaking injury are almost always under 5 years old and usually under 12 months old. When a reliable history is available, it typically involves a parent or other caregiver who shook an inconsolably crying baby out of anger and frustration. Often, however, the only information provided is that the child's mental status deteriorated or that seizures or respiratory difficulty developed. The involved caregiver may relate that an episode of relatively minor trauma occurred, such as a fall from a bed. Even without a supporting history, the diagnosis of Shaken Baby syndrome can still be made with confidence on the basis of characteristic clinical findings in the absence of a valid history or of an identified pathologic process that could present with similar signs and symptoms, such as a metabolic disease or clotting disorder. Such alternative diagnoses are usually readily identifiable by history, laboratory/radiologic testing, and clinical examination, including the appearance of the retina. Answers to important questions concerning the timing and circumstances of injury and the identity of the perpetrator sometimes cannot be inferred from medical evidence alone.

The infant's head is particularly vulnerable to the effects of repeated acceleration-deceleration because of its relatively large mass in relation to the body and poor stabilization by neck muscles. Intracranial injury in shaken infants frequently includes subdural hematoma, unilateral or bilateral over the cerebral convexities, or in the interhemispheric fissure. Displacement of the brain in relation to the skull and dura mater ruptures bridging vessels, leading to subdural hemorrhages. Evidence of subarachnoid bleeding may often be apparent. Although on initial scans the brain can be normal, in many cases cerebral parenchymal damage is manifest on neuroimaging, acutely as edema, ischemia, or contusion, and in later stages as atrophy. Repetitive stresses and strains within the brain cause direct brain damage, which is magnified by subsequent cerebral edema hypoxic-ischemic damage. Some authorities, citing the frequency with which shaken baby syndrome victims also show evidence of having received blows to the head, think that impact is an essential component of this syndrome, although in many cases, no sign of impact is found.

Ocular Involvement

The most common ocular manifestation of shaking injury, present in approximately 85% of cases, are retinal hemorrhages. The absence of retinal hemorrhage does not rule out abuse. Rarely, retinal hemorrhage can occur without intracranial hemorrhage. Preretinal, nerve fiber layer, deeper intraretinal (dot and blot), or subretinal localization may be seen. Hemorrhages tend to be concentrated in or near the posterior pole, but frequently are so extensive that they occupy nearly the entire fundus. Retinal hemorrhages in shaken infants resolve over a period ranging from several days, to 1-2 weeks, rarely to several months, depending upon location and severity. For example, superficial flame hemorrhages often resolve quickly, whereas preretinal and subretinal hemorrhage take longer to resolve. Retinal hemorrhages cannot be dated with any precision. Vitreous hemorrhage can also develop, usually as a secondary phenomenon resulting from migration of blood that was initially intraretinal or preretinal. Occasionally, the vitreous becomes almost completely opacified by dispersed hemorrhage within a few days of injury. A vitrectomy should be considered if a vitreous hemorrhage does not clear as the development of amblyopia or other complication such as myopia are likely to result.

Some eyes of shaken infants show evidence of retinal tissue disruption in addition to hemorrhage. Full-thickness perimacular folds in the neurosensory retina, typically

Abusive Head Trauma/Shaken Baby Syndrome

with circumferential orientation around the macula that creates a crater-like appearance, are highly characteristic. Splitting of the retina (traumatic retinoschisis), either deep to the nerve fiber layer or superficial (involving only the internal limiting membrane), creates partially blood-filled cavities of considerable extent, usually in the macular region. Full-thickness retinal breaks and detachment are rare. Schisis cavities usually flatten out within a few weeks of injury, but can persist indefinitely. Although similar findings have been reported rarely in fatal crush injuries and fatal motor vehicle accidents, such histories are readily apparent and would allow rapid identification.

A striking feature of Shaken Baby syndrome is the frequent lack of external evidence of trauma. The ocular adnexa and anterior segments appear entirely normal. Occasionally, the trunk or extremities show bruises representing the imprint of the perpetrator's hands. In some cases, rib fractures or characteristic metaphyseal fractures of the long bones result from forces generated during grasping of a limb or the perpetrator's hands encircling the thorax during the application of acceleration-deceleration forces. It must be kept in mind, however, that many shaken babies are also victims of other forms of abuse. Signs of impact to the head must be carefully sought, abdominal trauma specifically considered, and a radiologic skeletal survey performed in all cases of suspected abusive head trauma in this age range.

When extensive retinal hemorrhage accompanied by perimacular folds and schisis cavities are found in association with intracranial hemorrhage or other evidence of trauma to the brain in an infant without another clear explanation, abusive head trauma can be diagnosed with confidence regardless of other circumstances. Retinal hemorrhages without other ocular findings strongly suggests that intracranial injury has been caused by shaking, but alternative possibilities such as a coagulation disorder, normal birth hemorrhages, fulminant meningitis, and leukemia should be considered as well. Severe accidental head trauma (e.g. sustained in a fall from a second-story level or a motor vehicle collision) is infrequently accompanied by retinal hemorrhage, which is rarely extensive and usually confined to the posterior pole. Retinal hemorrhage is rare and has never been documented to be extensive following cardiopulmonary resuscitation by trained personnel. Terson syndrome appears to be rare in childhood and not likely to be associated with extensive multilayered retinal hemorrhage. Likewise, increased intracranial pressure or hypoxia are not known to be associated with extensive retinal hemorrhage. Papilledema is uncommon in Shaken Baby syndrome. Retinal hemorrhages resulting from birth trauma are common in newborns but do not persist beyond 1 month of age unless the hemorrhages are preretinal, subretinal or in the vitreous.

Currently, there is abundant evidence from multiple sources (perpetrator confessions, clinical studies, postmortem studies, mechanical models, animal models and finite element analysis) that repetitive acceleration-deceleration with or without head impact is injurious, and the primary cause of retinal hemorrhage in victims of *Shaken Baby syndrome* is vitreo-retinal traction. The well formed vitreous of infants and young children is very firmly attached to retinal blood vessels, the peripheral retina and the macula. Other factors such as increased intrathoracic pressure, increased intracranial pressure, brain trauma induced coagulopathy, hypoxia and anemia may have some role in modulating the appearance of retinal hemorrhage but are not likely to play a major pathogenic role, especially for extensive hemorrhage or schisis. Increased intracranial pressure, which can have many causes, does not lead to extensive retinal hemorrhages in the absence of abuse.

Abusive Head Trauma/Shaken Baby Syndrome

Ophthalmologists should be prepared to promptly respond to requests for consultation in cases of suspected abusive head injury. Ophthalmology consultations are appropriate when there is suspicion of abusive head injury based on other relevant findings or a history of witnessed events that might result in such injury. In addition, consultation may be appropriate for children who experience sudden unexplained life threatening events, including seizures and apnea. Even within the first 24 hours after injury hemorrhages may start to resolve or even worsen to some degree. Although photodocumentation is not required, it may later aid in providing recollection of the findings. Clinical notes that carefully detail (with or without hand drawn figures) the number, extent, pattern, type, and laterality of all ocular findings are essential. Findings should be communicated with the responsible pediatric child abuse teams that are found in many institutions. In the absence of such support, the ophthalmologist must honor their legal obligation to report a suspicion of child abuse via appropriate state or provincial reporting pathways. Protocols have been published for postmortem examination of the eyes and orbital tissues.

Prognosis

Up to one third of children with head trauma die from their injuries. Poor visual and pupillary response have been correlated with a higher risk of mortality. Survivors often suffer permanent impairment, ranging from severe retardation and quadriplegia to mild learning disability and motor disturbances. The most common cause of visual loss is cortical injury followed by optic atrophy. Dense vitreous hemorrhage, usually associated with deep traumatic retinoschisis, carries a poor prognosis for both vision and life.

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