# Abusive Head Trauma/Shaken Baby Syndrome

## Introduction

Although the majority of eye injuries in childhood are as a result of accidents, some 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 over 1 million confirmed victims per year in the United States alone.  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 physical 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 are inconsistent with the extent of injuries (e.g., bruises on multiple aspects of the head after a simple 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 fully 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 physical 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, are possible mechanisms of non-accidental 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 questionable, particularly in infants. Cataract or lens dislocation may be a sign of recent or previously inflicted injury or trauma. The 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. 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. The essential features were first identified in the early 1970s and it has become widely recognized as one of the most serious manifestations of physical child abuse.

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 spontaneously deteriorated or that seizures or respiratory difficulty developed. The involved caregiver may relate that an episode of relatively minor trauma occurred, such as a short fall. 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 or 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, and/or in the interhemispheric fissure. Displacement of the brain in relation to the skull and dura mater results in rupture of bridging vessels, leading to subdural hemorrhage. 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 and hypoxic-ischemic damage. Some authorities, citing the frequency where shaken baby syndrome victims also show evidence of having received blows to the head, think that impact is an essential component of this syndrome. However, in many cases, no sign of impact is found and perpetrators have confessed to shaking both with and without blunt head impact.

## Ocular Involvement
The most common ocular manifestation of a shaking injury, present in approximately 85% of cases, is retinal hemorrhages.  However, the absence of retinal hemorrhage does not rule out physical abuse. Rarely, retinal hemorrhage can even occur without intracranial hemorrhage. Preretinal, nerve fiber layer, deeper intraretinal (dot and blot), or subretinal hemorrhages 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 can 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 precision, especially by visual examination. Vitreous hemorrhage can develop as well, usually as a secondary phenomenon resulting from migration of blood that was initially beneath the internal limiting membrane, intraretinal or preretinal. Occasionally, the vitreous becomes almost completely opacified by dispersed hemorrhage within a few days of injury. A vitrectomy may be considered if a vitreous hemorrhage does not clear as the development of amblyopia or other complication, such as myopia, is a significant concern. Peripheral retinal non perfusion, detected by intravenous fluorescein angiography, has more recently been recognized as a possible outcome in shaken baby syndrome as well.

Some eyes of shaken infants show evidence of retinal tissue disruption in addition to hemorrhage. Full-thickness perimacular folds of the neurosensory retina, typically with circumferential orientation around the macula, creating a crater-like appearance, are highly characteristic of a shaking injury to the retina. 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, sometimes of considerable extent, usually in the macular region. Full-thickness retinal breaks, macular hole and detachment are rare but can occur. 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, an 11 meter fall, 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 of the eyes generally 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, 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.

Multiple studies have shown high specificity and odds ratios for child abuse in the presence of extensive multilayered retinal hemorrhages extending to the ora serrata, particularly when there are retinal folds, retinoschisis, and no other obvious explanatory history or disease. 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. The presence of retinal hemorrhages without other ocular findings strongly suggests that intracranial injury has been caused by repetitive acceleration-deceleration forces with or without blunt head impact, but alternative possibilities, such as a coagulation disorder, birth hemorrhages, fulminant purulent meningitis, ruptured aneurysm, and leukemia should be considered as well. Extensive retinal hemorrhages are rarely seen in critically ill patients in the absence of such easily recognized disorders. Severe accidental head trauma, even when 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 even by untrained personnel. Terson syndrome (intraocular hemorrhage resulting from a hyperacute rise in intracranial pressure as seen with a ruptured intracranial aneurysm or vascular anomaly) appears to be rare in childhood and not likely to be associated with extensive multilayered retinal hemorrhage except in the presence of hyperacute rises in intracranial pressure as seen in a ruptured aneurysm. Otherwise, increased intracranial pressure and factors, such as hypoxia, are not known to be associated with extensive retinal hemorrhage. Papilledema occurs in less than 10% of shaken baby syndrome victims. Retinal hemorrhages resulting from birth trauma are common in newborns, and may be extensive, but nerve fiber layer hemorrhages do not persist beyond 1 week and intraretinal hemorrhages do not persist beyond 3–4 weeks of age except for isolated dense hemorrhages, such as in the fovea. Birth induced preretinal, subretinal, or vitreous hemorrhages may last longer.

Currently, there is abundant evidence from multiple sources including perpetrator confessions, clinical studies, postmortem studies, mechanical models, animal models, and computer simulation 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.

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 injury. In addition, consultation may be appropriate for children who experience sudden unexplained life threatening events, including seizures and apnea. Retinal hemorrhages may start to resolve within the first 24 hours after injury or, even worsen, to some limited degree. Therefore, timely ophthalmological evaluation is important. Although photodocumentation is not required, it may later aid in providing recollection of the findings as well as enabling communication of the extent of retinal injuries to medical personnel and other authorities. Ancillary testing with

optical coherence tomography and intravenous fluorescein angiography may also be useful. Clinical notes are essential and should carefully detail the number, extent, pattern, type, and laterality of all ocular findings (with or without hand drawn figures). Findings should be communicated with the responsible pediatric child abuse teams 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 has been correlated with a higher risk of mortality. Survivors often suffer permanent impairment, ranging from severe mental retardation and quadriparesis 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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