CLINICAL REPORT Guidance for the Clinician in Rendering Pediatric Care

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The Eye Examination in the Evaluation of Child Abuse

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Reaffirmed With Reference & Data Updates

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Child abuse can cause injury to any part of the eye. The most common manifestations are retinal hemorrhages (RHs) in infants and young children with abusive head trauma (AHT). Although RHs are an important indicator of possible AHT, they are also found in other conditions. Distinguishing the number, type, location, and pattern of RHs is important in evaluating a differential diagnosis. Eye trauma can be seen in cases of physical abuse or AHT and may prompt referral for ophthalmologic assessment. Physicians have a responsibility to consider abuse in the differential diagnosis of pediatric eye trauma. Identification and documentation of inflicted ocular trauma requires a thorough examination by an ophthalmologist, including indirect ophthalmoscopy, most optimally through a dilated pupil, especially for the evaluation of possible RHs. An eye examination is helpful in detecting abnormalities that can help identify a medical or traumatic etiology for previously well young children who experience unexpected and unexplained mental status changes with no obvious cause, children with head trauma that results in significant intracranial hemorrhage and brain injury, and children with unexplained death.

INTRODUCTION

Direct or indirect trauma to the eye may be caused by child physical abuse. The most common manifestations are retinal hemorrhages (RHs), seen in approximately 75% of children who are victims of

abstract

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abusive head trauma (AHT), and are thought by many authors to be associated with the child experiencing repeated acceleration or deceleration forces with or without blunt force head impact.[1] Ophthalmologists are often asked to examine children when there is a concern of AHT. Such concern arises when there are signs of head and body injury, such as intracranial hemorrhage, but ocular injury is also a consideration when children present with unexplained alterations of consciousness or unexplained seizures. Ophthalmologists also have an independent duty to conduct a full eye examination when they have a concern about possible abuse as a cause of ocular injury. Ophthalmologists have an obligation to report their suspicions to the appropriate child protective service agency and, when available, consult with child abuse pediatricians or other professionals in the field of child abuse for guidance and additional assistance.

INDICATIONS FOR OPHTHALMOLOGY CONSULTATION

Eye abnormalities can be caused by direct blunt trauma or can be associated with more global brain injury. Children with inflicted injury to the eye come to the attention of the ophthalmologist in a number of ways. Most often, an ophthalmologist is consulted by colleagues because of a concern of AHT in a child with neurologic or other concerning injury. On occasion, an infant or child may present directly to an ophthalmologist with symptoms of neurologic or eye pathology that has resulted from an undisclosed inflicted injury. Examples include infants or children with an acquired strabismus from elevated intracranial pressure resulting from trauma or a child with a hyphema from an inflicted eye injury. Uncommonly, RHs may be discovered unexpectedly during a routine ophthalmologic examination. In children with AHT, the absence of visual compromise or the lack of external indicators of eye injury does not rule out the possibility of significant RHs. Therefore, one cannot rely on ocular signs or symptoms to determine which children might benefit from ophthalmologic consultation. Rather, ophthalmologic consultation is based on objective indicators of possible eye pathology, including findings that raise concern of possible AHT (eg, increased intracranial pressure, intracranial hemorrhage, or unexplained coma). An ophthalmologic examination is not an appropriate screening test for AHT in neurologically well-appearing infants who have an extracranial injury suggestive of abuse because RHs are uncommonly identified in this setting.[2,3]

The ophthalmologist is in a unique position to give a detailed clinical description of the patient's hemorrhagic retinopathy. The indirect ophthalmoscope provides a wide and stereoscopic field of view and enables the ophthalmologist to examine the anterior retinal edges (ora serrata), which is not possible using the direct ophthalmoscope even if the pupils are dilated. Eye examination for this purpose is best performed by an ophthalmologist. When indicated, a slit lamp inspection of the anterior segment to identify signs of trauma (eg, hyphema) can be helpful. Examination for an afferent pupillary defect (Marcus Gunn pupil) before pharmacologic pupillary dilation can be performed to identify optic nerve injury. Ophthalmologic consultation in the setting of suspected abuse is recommended for any child with visible injury to the eye, unexplained alterations of consciousness, intracranial hemorrhage, coagulopathy, or possible medical disease that might mimic abuse.[4]

THE EYE EXAMINATION

Although health care professionals other than ophthalmologists may be skilled at detecting the absence or presence of RHs,[5] a full view of the retina and characterization of the number, type, location, and pattern of the hemorrhages require consultation by an ophthalmologist by using indirect ophthalmoscopy, preferably with a dilated pupil. When there is a concern about transiently stopping pupillary reactivity, which affects the ability to monitor neurologic status, techniques such as dilation of 1 eye at a time, use of short-acting mydriatics, and use of a lens that affords some view through an undilated pupil can be used to allow indirect ophthalmoscopy, preferably within the first 24 hours and ideally within 72 hours after the child's acute presentation because some RHs resolve quickly. Even if the need for an eye examination is realized after 72 hours, ophthalmologic consultation may still be useful to identify persistent abnormalities, such as hemorrhages, retinoschisis, chorioretinal scars, and papilledema. Ophthalmologists can recognize characteristic findings of diseases of the eye and retina that are in the differential diagnosis of RHs, such as patterns suggestive of vasculitis, the peripapillary hemorrhages of papilledema, retinal infection, or leukemic infiltrates. Although not required, photodocumentation may enhance but not replace detailed written descriptions and drawings of eye findings when available. Regardless, a detailed documentation of the eye examination is essential.[6] Although several documentation and grading tools have been suggested, there is not yet a standard accepted tool.[7] Diagnostic impressions should objectively address the level of concern for abuse, relevant differential diagnosis, additional recommendations, and suggestions for further medical or ophthalmologic evaluation and possible reporting. As appropriate, acceptable terminology to describe the hemorrhages might include

"nonspecific" (with differential diagnosis), "suggestive," or "highly suggestive" of AHT. It may even be appropriate in some situations to indicate that there is no other reported cause of the retinal picture in the specific circumstance of historical and medical findings.

In recent years, researchers using techniques such as optical coherence tomography have documented the vitreoretinal interface and schisis cavities in victims with AHT.[8] Intravenous fluorescein angiography may detect characteristic areas of peripheral retinal nonperfusion, especially in the months after the incident.[9,10] These ancillary tests also have the potential to improve the understanding of pathophysiologic mechanisms of RHs and enable the ophthalmologist to identify retinal details not previously documented by clinical examination.

ORBITAL AND OCULAR INJURIES RESULTING FROM ABUSE

Injuries to the globe and surrounding structures are well described in child abuse and usually result from blunt trauma. Periorbital ecchymosis may result from abusive or accidental trauma to the forehead or periorbital tissues. Bilateral periorbital ecchymosis may be secondary to forehead trauma, basilar skull fractures, or subgaleal hematoma and can be seen in neuroblastoma and leukemia.[11] Frontal bone and orbital roof fractures are uncommon injuries in infants and young children and are even less common in those abused.[12,13] In children, these fractures are the result of significant blunt force injury and are best identified with computed tomography (CT), including a three-dimensional CT.[14]

Subconjunctival hemorrhages are well described in infants who are abused, who often but may not always have additional manifestations of abuse at the time of diagnosis.[15,16] Causes of subconjunctival hemorrhage in young children include direct eye trauma, indirect forces to the vessels from sudden increases in intrathoracic pressure, birth, or medical disease, including hemorrhagic conjunctivitis, pertussis, or hematologic disorders (thrombocytopenia in particular).

Traumatic hyphema can result from either blunt or penetrating trauma to the eye. In an older child, hyphema is commonly the result of an accidental high-energy blow to the globe rather than abuse.[17] Traumatic cataract may present in association with a hyphema at the time of injury or even weeks or months thereafter. Direct injury to the eye can also cause corneal abrasions or lacerations, globe rupture, or damage to the iris. Globe rupture may occur from blunt impact or contact with a sharp object. Any significant injury to the eye or orbit is best referred to an ophthalmologist for complete eye examination and management.

RHs AND AHT

RHs have been recognized as a key indicator of abusive head injury for more than 30 years.[18,19] Although mild and moderate RHs can be seen in a number of medical and traumatic conditions in children,[20] prospective and retrospective clinical studies have revealed that severe RH is strongly associated with AHT.[21,22] Approximately 25% of victims of AHT have no RH, and one-third of all cases have mild to moderate RH.[22] In 2 recent systematic reviews comprising over 30 clinical studies and thousands of children, the strong association of severe RH with AHT was confirmed.[1,23] In general, the number and severity of RHs correlate with the severity of neurologic injury.[24,25] For example, RHs are found infrequently in neurologically normal patients and are most frequently identified at autopsy in cases of fatal AHT. RHs in AHT can vary in number, size, and location within the retina. They can be bilateral or unilateral, confined to the peripapillary area or posterior pole, or can extend to the ora serrata. RHs can be subretinal, intraretinal, or preretinal and may extend into the vitreous.[1] Retinal folds, retinoschisis (splitting of the retinal layers), and retinal detachment (less common) are caused by trauma, although rare exceptions are reported.[26] RHs that are too numerous to count, are multilayered, bilateral, and extend to the ora serrata are highly specific for AHT.[1,27] Traumatic macular retinoschisis and folds are usually specific for AHT, although they have been reported after fatal accidental trauma, including head crush injuries, motor vehicle accidents, and falls.[1,28–31] Lesions similar to retinal folds and retinoschisis have been seen in leukemia, sickle cell anemia, and infantile aneurysm. In such cases, there may be isolated elevation of the internal limiting membrane of the retina with sub–internal limiting membrane blood, usually without surrounding retinal folds. These entities are readily excluded from the differential diagnosis by history, epidemiologic factors, or diagnostic testing. There are many causes of retinal detachment, which is uncommon in AHT; history and examination along with diagnostic testing assist in the differential diagnosis. On occasion, traumatic RH can occur in the absence of intracranial injury identified by imaging.[32,33]

Postnatal traumatic RHs cannot be accurately dated. One recent study reveals that intra-RHs resolve more quickly than pre-RHs, and hemorrhages that are too numerous to count only last for a few days after injury.[34] For example, these data reveal that the presence of pre-RHs without intra-RHs indicates an injury of at least a few days to 1 week in age. Although 1 article revealed that RH may worsen after admission,[35] this was not observed in another study.[36]

The degree of RH worsening after admission would not turn a mild retinopathy into severe RH, retinoschisis, or retinal folds.

RHs IN OTHER DISEASES

Although extensive, multilayered RHs that extend to the ora serrata, macular retinoschisis, and retinal folds are each highly suggestive of AHT, the diagnosis of AHT is not made on the basis of eye examination alone. History, physical examination, and diagnostic testing are always critical in establishing and working through a reasonable differential diagnosis. RHs have other etiologies, especially in critically ill children, including meningitis, leukemia, coagulopathy, and retinal disorders, all of which are identified by history, general and ocular examination, and appropriate medical evaluation. RHs in these diagnoses are almost always few in number and are confined to the posterior retina, although exceptions exist.[20,37] Hyperacute elevations of intracranial pressure, as seen in crush injuries or ruptured aneurysms, can also occasionally result in severe RHs.[38] Occipital impact or epidural hemorrhage may lead to RH and is usually intraretinal and confined to the posterior pole.[39,40]

The birth process often results in RHs, in part influenced by the delivery method. RHs that result from birth are most common after vacuum-assisted vaginal delivery (>70%) and are least common after routine cesarean delivery (<20%).[41,42] Severe RHs, macular retinoschisis, and retinal folds have not been reported in association with birth. Timing of birth RHs to resolution is well established, with flame hemorrhages resolving by 2 weeks (usually by 72 hours) and dot or blot hemorrhages by 6 weeks (but usually by 2 weeks).[43–45] Pre-RHs, foveal RHs, and the rarely present sub-RHs that result from birth may take many weeks or several months to resorb.[41]

Findings of systemic illness can be identified in an ocular fundus examination and may shed light on the etiology of a child's symptoms, such as necrotic retinitis from infection, retinal vascular abnormalities in Menkes disease, papilledema, or retinal manifestations of leukemia or bacterial endocarditis. There are no retinal ocular findings in sudden infant death syndrome, although routine ocular examination has not been common practice in these cases.[6,46] Perhaps such eye examinations would be useful, but further study is needed. Studies have revealed that seizures, cough, apnea, and childhood vaccinations do not cause RH.[47–49] Elevated intracranial pressure, except when hyperacute (eg, ruptured aneurysm, severe sudden head injury), does not result in extensive RH beyond the peripapillary area nor macular retinoschisis or folds.[38,50]

Excluding RHs that are associated with birth, AHT is the leading cause of RHs in infants. Victims of AHT present to medical care with a wide range of symptoms, from mild irritability and vomiting to unexplained coma or seizures. Some present with a false history of trauma (most often a short fall), and others present with only the symptoms that resulted from their abuse. Unsuspecting physicians misdiagnose up to one-third of victims with AHT presenting symptoms depending on the child's age, severity of symptoms, and family composition.[51] A retinal examination is not a substitute for brain imaging when assessing infants who have been physically abused who have no neurologic symptoms of AHT.[2]

MECHANISMS AND PATHOPHYSIOLOGY OF RHs

The pathogenesis of RHs is multifactorial. In recent years, important trends have emerged in the understanding of RHs attributable to AHT in young children.[52] Evidence has continued to increase in support of the diagnostic specificity of severe RH as an indicator of AHT in children with and without evidence of blunt impact injury to the head.[1,21,53] Extensive literature, including clinical studies and computer modeling, support this observation.[54,55] This research, along with clinical experience, support the role of vitreoretinal traction sustained during the repetitive acceleration or deceleration mechanism that characterizes shaking as an important contributory factor in causing RH and macular retinoschisis.[6,8,54,56] Although the abusive traumatic event is the primary etiology of the RH, factors such as hypoxia, anemia, reperfusion, autonomic vascular dysregulation, significant shifts in sodium balance, coagulopathy, and intracranial pressure elevation may modulate the appearance of RHs.[57] Further research is needed to better define the role of these and other factors as the understanding of the pathophysiology and diagnostic specificity of RH continues to evolve.

OUTCOMES

Visual morbidity from physical child abuse is significant, affecting more than 40% of children with severe AHT.[58] In AHT, visual outcome is not usually related to direct ocular trauma but is instead related to occipital cortical damage and/or optic nerve injury.[59] RHs generally resolve without sequelae and, depending on the type of hemorrhage, can resolve quickly.[34] Retinal causes of visual loss from AHT include retinal detachment, macular scarring or fibrosis, and vitreous hemorrhage, which can lead to amblyopia if the hemorrhage does not resorb without surgery. Infants and children with ocular injury from any cause require careful ophthalmologic follow-up to maximize their visual outcome.

THE POSTMORTEM EYE EXAMINATION

An autopsy is a unique opportunity for examination not only of the eye and its contents but also of the orbital tissues, which may yield findings helpful in determining the cause of the child's death. Although ophthalmology consultation can be obtained after death and before an autopsy and the use of monocular indirect ophthalmoscopy has been reported by nonophthalmologists,[60] proper autopsy techniques provide a more accurate assessment of ocular and retinal findings. Even when premortem ophthalmoscopy is performed, postmortem examination is necessary to view the orbital tissues. When possible, an examination by a trained ocular pathologist is ideal. Postmortem eye and orbital tissue examination is another means of documenting RH and retinoschisis but may also reveal hemosiderin deposition from previous events, such as hemorrhages into fat, muscles, or cranial nerve sheaths as well as intradural optic nerve sheath hemorrhages, all of which have diagnostic significance in identifying abused children.[61]

One obstacle to postmortem examination of the eyes and orbits has been a societal distaste or resistance, which in some cases has led to fear among pathologists of legal repercussion.[62] This may reflect a cultural or emotional objection specifically to eye removal. There might be a misconception that eye or orbital removal will alter the appearance of the body postmortem at a funeral viewing when in fact this is not the case. Techniques are now detailed to allow for proper removal of the eye and orbits without disfigurement. En bloc removal of the globe with all orbital tissues is recommended.[62] Although consent is not routinely obtained for coroner cases or forensic autopsies, there may be situations or jurisdictions in which specific consent for eye and orbital tissue removal is required. If a parent or guardian refuses this procedure, it may be necessary to seek legal intervention, such as a court order or intervention by child protective services, to allow the procedure to be performed.

CHALLENGES AND FUTURE DIRECTIONS

Although a retinal examination may assist in identifying the cause of unexplained mental status changes, seizures, intracranial hemorrhage, or unexplained early childhood deaths, premortem clinical ophthalmologic consultation and postmortem removal of the eyes and orbital tissues are not routine practices in some centers. Failure to conduct these procedures, particularly when there is no other explanation for a life-threatening event or death, risks losing an important opportunity to gain valuable information. Information gained in such an evaluation might lead to identifying an etiology and in the case of a surviving child, prevent death by identifying potential risks of abuse or recognizing other diseases.

There continue to be reports of novel, unsubstantiated causes of RH and other ocular findings in young children, such as vitamin D deficiency or vaccination.[63] Although there is no credible evidence that vitamin D deficiency or routine childhood vaccinations cause RH,[48] practitioners should be open to reports that broaden and at the same time narrow the RH differential diagnosis. For example, mild posterior pole RHs now have been described in osteogenesis imperfecta.[64] Cardiopulmonary resuscitation is not a cause of widespread RH.[65,66,67] Only with more widespread use of ophthalmologic consultation and postmortem ocular and orbital examination can such possibilities be discovered or refuted and then appropriately fit into the differential diagnostic process.

Photodocumentation and other emerging technologies, when available, have also proven to be potentially important ophthalmic procedures in documenting retinal abnormalities for both clinical and educational purposes.[68] Further research regarding these new technologies will lead to advances in the understanding of RHs and improve clinical evaluation.

GUIDANCE FOR PHYSICIANS

Ophthalmologic consultation is an important part of the diagnostic evaluation of all previously well children younger than 5 years old who experience unexplained coma, seizures, intracranial hemorrhage or injury, or have a systemic disorder known to have ocular manifestations. A complete eye examination is also important in the evaluation of unexplained death in infants and children. As mandated reporters, all physicians have a legal and ethical duty to report suspected abuse to local or state child protective service agencies. Reports can be made by calling their state's toll-free child abuse reporting hotline. Whenever possible, the accompanying parent(s) or guardian(s) should be notified about the concern and the need to report. It can be helpful to raise concern about the finding and, while not apportioning blame, inform the family that because of the nature and circumstance of the examination findings, a report for further investigation is mandated by law and serves to prevent the child from being injured again should that prove to be the cause of the observed findings. Additional guidance about reporting is available from the American Academy of Pediatrics.[69]

Specific guidance for the eye examination in the evaluation of child abuse is as follows:

- Retinal examination by an ophthalmologist is particularly important when there is a suspicion of AHT, although RHs may occasionally be found in infants and children with other causes of intracranial hemorrhage. Multiple (too numerous to count), bilateral, and multilayered hemorrhages that extend to the periphery of the retina are highly specific for AHT, and when identified, a complete evaluation for additional injuries is warranted.
- Full indirect ophthalmoscopic examination through a dilated pupil is the optimal method of examining the retina in these circumstances. Examiners should include a detailed description of findings, such as the number, type, location, and pattern of RHs when present. Retinal abnormalities may be photographed after the clinical examination when a camera is available. When indicated, a slit lamp inspection of the anterior segment to identify signs of trauma (eg, hyphema) can be helpful. Examinations for an afferent pupillary defect (Marcus Gunn pupil) before pharmacologic pupillary dilation should be performed to identify possible optic nerve injury.
- Because findings such as RH may be transient, it is desirable that the ophthalmologic consultation take place within 24 hours of the patient's presentation for medical care but certainly within 72 hours.
- In potential victims of abuse who are neurologically asymptomatic, a retinal examination is not an appropriate screening test for brain injury; rather, such children should undergo brain imaging (such as an MRI or CT) as the appropriate screen if indicated. If the neuroimaging result is normal, yet concern still exists about possible AHT, then eye examination may still be considered.
- Pediatricians who suspect a diagnosis of AHT should make a prompt referral to an ophthalmologist, who should be available for prompt evaluation of the patient because important safety and medical issues are dependent on determining an appropriate diagnosis.
- When pharmacologic pupillary dilation is believed to be undesirable, as in children with severe central nervous system injury, timely ophthalmologic consultation is still helpful. An attempt to view the retina and optic nerve through the use of direct ophthalmoscopy, small pupil indirect ophthalmoscopic techniques, sequential pharmacologic dilation, and/or fast-acting mydriatics (eg, phenylephrine 2.5%) can still yield important information.
- When a previously well child younger than 5 years old dies without explanation, regardless of whether a premortem retinal examination was conducted, examination of the eyes and orbital tissues as part of the autopsy may shed light on the cause of death. When possible, an ocular pathologist should perform the examination.
- Postmortem eye removal is not necessary in children who have clearly died of witnessed severe accidental head trauma or otherwise readily diagnosed systemic medical conditions.

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ABBREVIATIONS

AHT: abusive head trauma
CT: computed tomography
RH: retinal hemorrhage

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