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Clinical Report—The Eye Examination in the Evaluation of Child Abuse

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abstract

FREE

Retinal hemorrhage is an important indicator of possible abusive head trauma, but it is also found in a number of other conditions. Distinguishing the type, number, and pattern of retinal hemorrhages may be helpful in establishing a differential diagnosis. Identification of ocular abnormalities requires a full retinal examination by an ophthalmologist using indirect ophthalmoscopy through a pupil that has been pharmacologically dilated. At autopsy, removal of the eyes and orbital tissues may also reveal abnormalities not discovered before death. In previously well young children who experience unexpected apparent life-threatening events with no obvious cause, children with head trauma that results in significant intracranial hemorrhage and brain injury, victims of abusive head trauma, and children with unexplained death, premortem clinical eye examination and postmortem examination of the eyes and orbits may be helpful in detecting abnormalities that can help establish the underlying etiology. *Pediatrics* 2010;126:376–380

BACKGROUND

When a previously well child experiences an apparent life-threatening event (ALTE) or unexpected death without obvious cause, pediatricians and other physicians must attempt to identify the etiology. In the case of an ALTE, one should consider diagnoses such as gastroesophageal reflux, seizures, other central nervous system disease, metabolic disease, breath-holding, and abusive head trauma (AHT). Retinal examinations have been used with limited success for screening ALTE victims for possible AHT.^{1,2} Victims of AHT present to medical care with a wide range of symptoms, from mild irritability and vomiting to unexplained coma or seizures.³ It has been estimated that approximately 4% to 6% of abused children present first to an ophthalmologist,⁴ and the most common ocular manifestation of abuse is retinal hemorrhage. Some children present with a false history of trauma, and others present with only the symptoms that resulted from their abuse. Unsuspecting physicians misdiagnose the condition of up to one-third of symptomatic victims, depending on their age, severity of symptoms, and family composition.⁵ When a child dies unexpectedly, considerations include previously undiagnosed or new systemic disease, sudden infant death syndrome, and covert abusive injury.

Retinal hemorrhages have been recognized as a key indicator of abusive head injury for more than 30 years, particularly in association with severe repetitive acceleration-deceleration forces with or without blunt head impact, in children younger than 5 years.^{6,7} Because retinal

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KEY WORDS

eye examination, child abuse, abusive head trauma, retinal hemorrhage

ABBREVIATIONS

ALTE—apparent life-threatening event

AHT—abusive head trauma

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The guidance in this report does not indicate an exclusive course of treatment or serve as a standard of medical care. Variations, taking into account individual circumstances, may be appropriate.

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hemorrhage rarely results in visual compromise, there may be no external indicators of eye injury unless the vision is significantly impaired by damage to the optic nerve or occipital cortex or there is retinal detachment, significant vitreous hemorrhage, or other severe disruption of the intraocular contents. Both eyes need to have significant visual compromise for a child to become visually symptomatic. Therefore, one cannot rely on ocular signs or symptoms to determine which children might benefit from ophthalmologic consultation and postmortem eye removal. Likewise, retinal examinations should not be limited to patients who are victims of suspected AHT. Searching for retinal hemorrhages as diagnostic criteria for AHT only in infants with suspected abuse creates a selection bias.

Although health care professionals other than ophthalmologists may be skilled at detecting the absence or presence of retinal hemorrhage,⁸ a full view of the retina and characterization of the number, types, and patterns of the hemorrhages requires consultation by an ophthalmologist using indirect ophthalmoscopy, preferably with a dilated pupil. Even when there may be a concern about transiently obliterating pupillary reactivity in the face of a need to monitor neurologic status acutely, 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 employed to allow indirect ophthalmoscopy, preferably within the first 24 hours and ideally within 72 hours after the child's acute presentation. Even if the need for eye examination is realized after 72 hours, ophthalmologic consultation may still be useful to identify persistent abnormalities such as hemorrhages, retinoschisis, and papilledema.

The range of retinal hemorrhage findings in AHT and accidental trauma is broad, and the severity of the findings can be informative of etiology. For example, retinal hemorrhage—predominantly intraretinal, in small numbers, and confined to the posterior pole of the retina—can be seen after significant accidental head injury or in AHT.^{6,9} More dramatic retinal hemorrhages—multilayered, too numerous to count, and extending to the edge of the retina (ora serrata)—can be seen after AHT, normal birth, fatal extreme accidental injury (such as motor vehicle accidents), and perhaps after fatal head crush injury.^{6,10–13} Ocular fundus examination can also reveal findings of systemic illness that may shed light on the etiology of a child's symptoms, such as a cherry-red spot in metabolic disease, retinal vascular abnormalities in Menkes disease, papilledema, and retinal manifestations of leukemia or bacterial endocarditis. There are no known retinal ocular findings in sudden infant death syndrome, although routine ocular examination has not been common practice in these cases.^{6,14}

Excluding retinal hemorrhages that are associated with vaginal delivery, AHT is the leading cause of retinal hemorrhages in infants. The association of retinal hemorrhage and AHT has been demonstrated repeatedly in clinical studies.^{15–17} Although retinal hemorrhages in AHT can be unilateral or bilateral and vary in degree, the severity of retinal hemorrhage often parallels the severity of brain injury.^{18,19} Because intraretinal hemorrhages may resolve quickly, a retinal examination is not a substitute for brain imaging when screening physically abused infants who have no neurologic symptoms for AHT.²⁰

The ophthalmologist is in a unique position to detail the hemorrhagic retinopathy. The indirect ophthalmoscope

provides a wide and stereoscopic field of view and enables the ophthalmologist to examine the anterior aspects of the retina to the ora serrata, which is not possible using a direct ophthalmoscope even if the pupils are dilated. Eye examination for this purpose should be performed by an ophthalmologist. Attention must be paid to special features, such as the presence of traumatic macular retinoschisis, because these features may have particular diagnostic significance for abuse.

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 differential diagnosis. This is particularly true when a child dies before clinical ophthalmologic consultation can be obtained. Even when premortem ophthalmoscopy is performed, postmortem examination is necessary for viewing the orbital tissues. When possible, examination by a trained ocular pathologist or ophthalmologist with experience in interpreting ocular pathology is ideal. Postmortem eye and orbital tissue examination is another means of documenting retinal hemorrhage and retinoschisis but may also reveal hemosiderin deposition from previous events and orbital findings, such as hemorrhage into the fat, muscles, and cranial nerve sheaths as well as intradural hemorrhage, all of which may have diagnostic significance in identifying abused children.²¹

One obstacle to postmortem examination of the eyes and orbits has been a societal distaste or resistance that, in some cases, has led to fear among pathologists of legal repercussion. 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 this is not, in fact, the case. Techniques allow

for proper removal of the eye and orbits without disfigurement.⁶ Although consent is not routinely obtained for coroner cases/forensic autopsies, there may be situations or jurisdictions in which specific consent for eye and orbital tissue removal may be sought. If a substitute decision-maker refuses this procedure, it may be necessary to seek legal intervention to allow the procedure to be performed.

STATEMENT OF THE PROBLEM

Although a retinal examination may suggest the etiology for ALTEs and previously unexplained early childhood deaths, premortem clinical ophthalmologic consultation and postmortem removal of the eyes and orbital tissues are not routine practice in some centers. Those who fail to conduct these procedures, particularly when there is no other explanation for the life-threatening event or death, risk 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 preventing further abuse or recognizing other disease.

In recent years, 3 important trends have emerged in the understanding of hemorrhagic retinopathy in young children. First, there has been a continued and dramatic increase in the evidence supporting the diagnostic specificity of severe hemorrhagic retinopathy as an indicator of AHT, in particular, severe repeated acceleration-deceleration with or without blunt head impact. Extensive literature worldwide, including clinical studies,⁷ animal models,²² and computer modeling,^{23,24} well beyond the scope of this report, support this theory. It seems that vitreoretinal traction and orbital injury sustained during the unique repetitive acceleration-

deceleration mechanism that distinguishes this form of abuse from single-impact trauma is the critical factor in causing retinal hemorrhage.⁸ Factors such as hypoxia, anemia, and intracranial pressure may play important secondary roles in modulating the appearance of retinal hemorrhages but do not, in and of themselves, result in such a retinopathy.⁶ Further research is needed to better define the role of these and other factors as our understanding of the pathophysiology and diagnostic specificity of retinal hemorrhage continues to evolve.

Second, there has been increasing recognition of the importance of detailing the appearance of the hemorrhagic retinopathy rather than simply using the term "retinal hemorrhage" generically. The differential diagnosis of a few intraretinal hemorrhages surrounding the optic nerve is vastly greater than that for "too-numerous-to-count" multilayered retinal hemorrhages that extend to the ora serrata.

Third, there continue to be novel reports of causes of retinal hemorrhage and other ocular findings in young children. These hypothesis-generating observations allow practitioners to broaden and at the same time narrow differential diagnosis. For example, mild posterior pole retinal hemorrhages have been described in osteogenesis imperfecta²⁵ as well as head trauma sufficient to cause epidural hemorrhage,⁹ and more severe hemorrhage was observed in 2 cases of fatal head crush injury.^{10,11} Only with more widespread use of ophthalmologic consultation and postmortem ocular and orbital examination can such entities be discovered and fit into the differential diagnostic process appropriately. Photodocumentation, when available, has also proven to be an important ophthalmic procedure for documenting retinal abnormalities for both clinical and educational purposes.

Full ophthalmic examination by an ophthalmologist and/or postmortem eye removal and pathologic examination can be a critical part of the diagnostic evaluation of previously well children who have experienced an unexplained ALTE or death and also for children who have disorders in which there are known associations with ophthalmic findings, particularly AHT.

GUIDANCE FOR PEDIATRICIANS

Ophthalmologic consultation should be part of the diagnostic evaluation of all previously well children younger than 5 years who experience an unexplained death or unexplained ALTE or have a systemic disorder known to have ocular manifestations.

- All infants and young children who present with significant intracranial hemorrhage should have an ophthalmologic consultation. Retinal examination is particularly important when there is a suspicion of AHT, but some retinal hemorrhages may occasionally also be found in infants and children with other causes of intracranial hemorrhage.
- Ophthalmologic examination should include, whenever possible, full indirect ophthalmoscopic examination through a dilated pupil. Descriptions of findings should be detailed and include the number, type, extent, and pattern of retinal hemorrhages, if present. Retinal abnormalities may be photographed after the clinical examination when a camera is available. When indicated, the examination should include slit-lamp inspection of the anterior segment to identify signs of trauma (eg, hyphema).
- Because findings such as retinal hemorrhage may be transient, it is desirable that the ophthalmologic consultation take place preferably

within 24 hours after the patient presents for medical care and ideally within 72 hours.

- A retinal examination is not an appropriate screening test for brain injury in neurologically asymptomatic potential victims of abuse. Such children should undergo brain imaging as the appropriate screen.
- When pharmacologic dilation is felt to be undesirable, as for children with severe unstable central nervous system injury, timely ophthalmologic consultation is still needed. An attempt should still be made 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%).
- When a previously well child younger than 5 years dies without explanation, regardless of whether a premortem retinal examination

was conducted, the eyes and orbital tissues should be removed en bloc at autopsy per published techniques.²⁶ When possible, an examination by an ocular pathologist or ophthalmologist with experience in interpreting ocular pathology is preferable.

- Postmortem eye removal may not be indicated in children who have clearly died from witnessed severe accidental head trauma or otherwise readily diagnosed systemic medical conditions.
- Ophthalmologic examination and/or postmortem eye and orbital tissue removal should be performed in all cases in which a child is alleged to have suffered significant morbidity secondary to a short fall or other minor trauma disproportionate to the clinical injury and consistent with child abuse.

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An error occurred in this American Academy of Pediatrics clinical report (doi: 10.1542/peds.2010-1397). The name of George S. Ellis, Jr, MD, Immediate Past Chairperson of the Section on Ophthalmology Executive Committee, was inadvertently omitted. The Academy regrets the error.

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Husson B, Hertz-Pannier L, Renaud C, et al. Motor Outcomes After Neonatal Arterial Ischemic Stroke Related to Early MRI Data in a Prospective Study. *Pediatrics*. 2010;126(4):e912–e918

An error occurred in this article by Husson et al (doi:10.1542/peds.2009-3611). On page e917, under the heading Acknowledgments, on line 4, this reads: “. . . , the Motorola foundation, . . .” This should have read: “. . . , La Fondation Motrice, . . .”

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