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SPECIAL ISSUE: ABUSIVE HEAD TRAUMA

The eye in child abuse: Key points on retinal hemorrhages and abusive head trauma

Gil Binenbaum · Brian J. Forbes

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Abstract This review presents an up-to-date overview of ocular injuries resulting from child abuse, with a spotlight on abusive head trauma. Retinal hemorrhage is a principle finding of inflicted head trauma. The specific pattern of hemorrhages holds valuable diagnostic information, which can help to guide multidisciplinary assessments of the likelihood of abuse. Indirect ophthalmoscopy through dilated pupils by an ophthalmologist is necessary for adequate examination and documentation of retinal findings. Initial pediatrician evaluation of the eye and indications for ophthalmological consultation are reviewed. Focus is then placed upon understanding retinal hemorrhage patterns, their diagnostic significance and likely pathophysiological mechanisms. The differential diagnosis of retinal hemorrhage in young children is discussed, highlighting key distinctions among retinal hemorrhage patterns, severity and frequencies, as well as other ocular findings. The most common cause of retinal hemorrhage in an infant is trauma, and most other causes can be identified by considering the hemorrhage pattern, ocular or systemic signs and the results of laboratory and imaging tests, when indicated.

Keywords Retinal hemorrhage · Abusive head trauma · Child abuse · Ocular injury · Shaken baby syndrome

G. Binenbaum · B. J. Forbes
The Children's Hospital of Philadelphia, The Perelman School of Medicine at the University of Pennsylvania,
Philadelphia, PA, USA

G. Binenbaum (✉) · B. J. Forbes
Pediatric Ophthalmology, 9-MAIN,
The Children's Hospital of Philadelphia,
34th Street and Civic Center Boulevard,
Philadelphia, PA 19104, USA
e-mail: binenbaum@email.chop.edu

Introduction

The eye can be a direct or indirect target of an inflicted injury. Ocular findings can provide valuable diagnostic information for the multidisciplinary team, particularly when there are limited external signs of abuse. This review provides an overview of the relevant eye examination and focuses upon retinal findings in abusive head trauma, including description, mechanism, differential diagnosis and diagnostic significance. With any form of head injury, the eyes should always be carefully examined. An ophthalmologist should be consulted if there are external signs of periorbital or ocular trauma or if there is a concern for abusive head trauma, for example when intracranial hemorrhage is present on neuroimaging. Examination of the eyes should be approached in a systematic fashion, inspecting each anatomical structure with adequate light and magnification. Adequate examination of the posterior structures of the eye requires a dilated fundus examination with indirect ophthalmoscopy to be performed by an ophthalmologist.

Anterior segment examination

The conjunctiva is a transparent mucous membrane that covers the sclera and inside of the eyelids. Subconjunctival hemorrhage or conjunctival laceration may indicate direct ocular trauma. Subconjunctival hemorrhage can also occur as a result of a sudden increase in intrathoracic pressure from blunt trauma to the thorax, severe vomiting, valsalva maneuver or paroxysmal coughing, a possibility that is increased in the presence of a coagulopathy. Abuse should be suspected if a child has subconjunctival hemorrhage with no physiological reason or known accidental trauma involving compression of the chest. The cornea is a clear, dome-shaped, transparent structure overlying the anterior chamber and iris. The cornea should normally be clear, and any focal or diffuse opacity or haze requires an evaluation by an ophthalmologist. A direct injury to the eye may cause a corneal abrasion or laceration, which can be very painful. Abrasions can be identified by applying fluorescein dye and shining a cobalt blue light from the direct ophthalmoscope or slit-lamp biomicroscope. Corneal or scleral lacerations from sharp objects or globe rupture from blunt trauma may be signs of an abusive injury. Such injuries are surgical emergencies and may result in long-term scar formation and irregular refractive errors (astigmatism), both of which could lead to vision loss. Breaks in Descemet's membrane (the endothelial basement membrane on the inner surface of the cornea) usually result from blunt or penetrating trauma and can also induce astigmatism and scarring. Trauma is the most common cause of a hyphema, or blood in the anterior chamber of the eye, located behind the cornea and in front of the iris. Blunt trauma to the head or the eye can cause a crystalline lens dislocation or opacity (cataract), often in association with a hyphema or periorbital ecchymosis and subconjunctival hemorrhage, and lead to vision loss and need for surgical intervention.

For anterior segment injuries, a much more detailed examination is obtained through the use of a slit-lamp biomicroscope, which provides an excellent magnified view of the eye and permits identification of visually and diagnostically significant findings otherwise too subtle to identify. Such instrumentation is necessary to diagnose intraocular inflammation and hemorrhage and to carefully examine the cornea and other structures. When anterior segment injury is suspected, ophthalmological consultation provides this more detailed examination.

Posterior segment examination

The retina covers a large area, extending from the optic nerve head and posterior pole to its termination at the ora serrata in the front third of the eye. Traditionally, most physicians use the direct ophthalmoscope to examine the posterior ocular structures, but even through dilated pupils, this instrument provides an inadequate retinal examination. The indirect ophthalmoscope, a head-mounted unit used in conjunction with a handheld condensing lens, provides a much wider-angle binocular view that simultaneously allows visualization of nearly the entirety of the posterior pole, which includes the optic disc and macula. By directing the view toward different parts of the fundus, most of the peripheral retina can be seen as well. Scleral depression, a technique in which the wall of the eye is manually compressed, can be used to bring the far peripheral retina into view. In an uncooperative child who is moving his or her eyes, fundus images may flash past the viewer. The much larger field of view provided by the indirect ophthalmoscope makes it more likely that important findings will come into view and generally allows a superior examination even in uncooperative children. Dilation of the pupils provides a significantly better view for the examiner, facilitating detection of retinal hemorrhage and permitting fundus photography. Pupillary constriction, which may result from the administration of barbiturates for sedation, can limit the yield of the examination, even with an indirect ophthalmoscope. The benefits of pharmacological dilation usually outweigh the loss of ability to monitor pupillary reaction for a few hours, but even when dilation is not possible, indirect ophthalmoscopy should be attempted and the results documented.

Generally, an ophthalmological consultation should be obtained to perform an adequate retinal examination. In one retrospective case series of 123 children admitted for subdural hematomas caused by abuse, non-ophthalmologists failed to detect retinal hemorrhages in 29% of affected children [1]. An ophthalmologist, who is adept with the indirect ophthalmoscope and more familiar with types and patterns of retinal pathology associated with conditions in the differential diagnosis, cannot only better detect findings but also provide a more knowledgeable opinion regarding the etiology of the findings and more detailed written and photographical documentation for medicolegal purposes.

Postmortem examination of the eyes and orbits can reveal signs of trauma and should be performed in cases of suspected abuse in infants and young children [2–5]. Both optic nerve sheath and intraocular hemorrhage are frequently reported findings in abusive head trauma victims. Nerve sheath hemorrhage tends to be most prominent anteriorly, may not extend the length of the optic nerve and frequently involves multiple layers, but it often shows a preponderance for the subdural space [6, 7]. Other evidence of acceleration-deceleration injury may include hemorrhage in the orbital fat or extraocular muscles [4, 5]. Protocols for forensic ophthalmic autopsy are available and should include gross photography of the open fixed globe, orbital exenteration and microscopic examination, ideally by an ocular pathologist.

Ophthalmological findings of abusive head trauma

The principal ophthalmological finding of abusive head trauma is retinal hemorrhage, which is seen in about three-quarters of cases, with a range of 50-100% depending upon the published series [3, 8–12]. The frequency of retinal hemorrhage is highest in autopsy cases and lowest in neurologically normal survivors, and the severity of retinal hemorrhage is associated with severity of neurological injury [13, 14]. Typically, hemorrhages are present in both eyes, although marked asymmetry or unilaterality is well recognized and does not alter the diagnostic significance. Correlation between laterality of intracranial findings (extra-axial hemorrhage, edema) and laterality of retinal findings has not been consistently found [10, 12].

The pattern of retinal hemorrhage is important diagnostically, as discussed below. Significant characteristics include the number, type and location of retinal hemorrhage, as well as the presence of retinoschisis and retinal folds. Types of retinal hemorrhage include intraretinal, preretinal and subretinal. Vitreous hemorrhage can be seen as well. Locations of the retinal hemorrhage include peripapillary (around the optic nerve), posterior pole (perivascular or along the retinal vessel arcades, and in the macula), retinal periphery and ora serrata (anterior extent of the retina). Retinal hemorrhage in abusive head traumas range from no retinal hemorrhage to mild retinal hemorrhage (few in number, exclusively intraretinal and confined to the posterior pole) to moderate or severe (multilayered and extending out to the ora serrata) (Fig. 1). Dense preretinal or vitreous hemorrhages may obscure underlying retinal hemorrhage.

Circular retinal folds are a distinctive but not pathognomonic finding in abusive head trauma, having also rarely been reported after severe direct head injury and with spontaneous subarachnoid hemorrhage in adults. A fold may be continuous or discontinuous, partial or complete. It may encompass just the macula or surround the entire posterior pole including the optic nerve. Both peripheral and macular traumatic retinoschisis (splitting of the retinal layers) are highly suggestive of abusive head trauma. Most commonly, only the internal limiting membrane and/or nerve fiber layer is pulled away by the vitreous, though deeper schisis and even focal macular detachment can also occur. The schisis cavity is often surrounded by a hypopigmented or hemorrhagic circumlinear line or elevated retinal fold (Fig. 2). Histologically, one often observes vitreous still attached at the edges of the lesion and depigmentation of the retinal pigmented epithelium underlying the folds. A hemorrhagic macular cyst may exist with dense hemorrhage within a schisis cavity, often with a meniscus demarcating red blood cells below from serum above. Blood may break through the schisis wall into the vitreous, obscuring the edge changes of a cavity, which may become apparent only when the blood resolves. Papilledema is found in a minority (~10%) of abusive head trauma patients [10–12].

Fig. 1 Color fundus photograph of the right eye of an infant who was a victim of abusive head trauma. Numerous intraretinal hemorrhages (arrows) are seen extending from the posterior pole into the retinal periphery. A preretinal hemorrhage (double headed arrow) is visible just inferior to the optic disc (D)

Documentation

Documentation of ophthalmic findings plays a key role in interpretation of findings by the multidisciplinary child abuse team. Many examiners may use quickly crafted drawings that may not adequately reflect the number, types and locations of retinal hemorrhage in each eye, as ophthalmologists are used to using such illustrations in the management of other retinal diseases. Detailed written descriptions using words in combination with carefully drawn schematic drawings are more useful. Retinal photography is very helpful for documenting retinal hemorrhages but requires expensive equipment not readily available in all centers. A well-documented clinical note is adequate and a lack of photographs should not be considered a flaw in the forensic evaluation of the child, as long as the hemorrhages are described well and detailed by other forms of documentation.

Diagnostic significance of retinal hemorrhages

Beyond the neonatal period, retinal hemorrhages are exceedingly uncommon in infants and young children. The most common cause of retinal hemorrhage is trauma in the form of birth "trauma" in newborns and abusive head trauma in infants and children [3, 15]. Both the presence and increasing severity of retinal hemorrhages are associated with increased odds of inflicted versus accidental injury in children with intracranial hemorrhage, especially in children younger than 6 months of age [8, 9, 16]. Retinal hemorrhages that are numerous, multilayered and widespread (extending from the posterior pole to the far retinal periphery) are highly suspicious for abusive head trauma, unless there is unambiguous severe accidental head trauma and/or confounding systemic disease. In 2010, Bhardwaj et al. [8] conducted a systematic review, using stringent selection criteria to identify studies with case definition provided by confession, third-party witness, legal decision or multidisciplinary assessment and free of circular logic. Among 560 abusive head trauma cases, retinal hemorrhage was found in 74% (range: 51-100%) of clinical exams and 82% (range: 63-100%) of autopsy cases. These figures represent the sensitivity of retinal hemorrhage for abusive head trauma. In contrast, retinal hemorrhages were present in only 6% of 242 accidental traumas from 4 prospective controlled studies, providing a specificity of retinal hemorrhages for abusive head trauma of 94% (range: 90-100%).

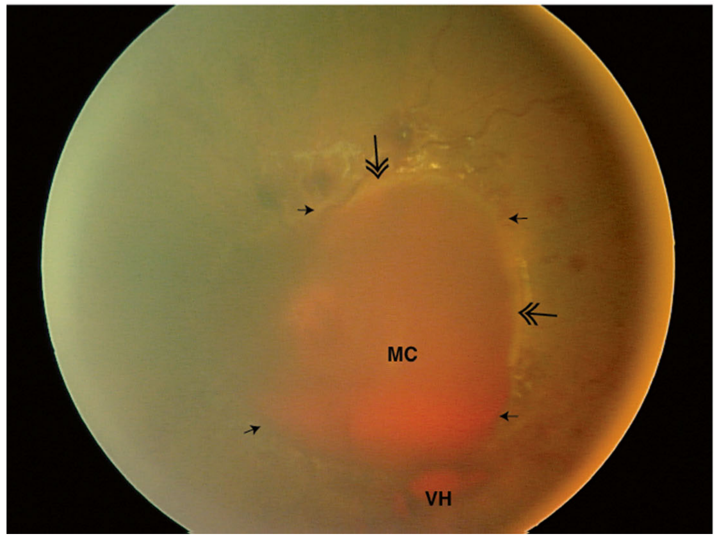

Fig. 2 Color fundus photograph of the left eye of an infant who suffered abusive head trauma. A hemorrhagic macular cyst (MC) is seen within an area of retinoschisis (bordered by arrows). Retinal folds (double headed arrows) are visible surrounding the borders of the cyst, and globules of vitreous hemorrhage (VH) are seen at the inferior edge of the cyst

The specificity increased with preretinal, peripheral and moderate to severe retinal hemorrhages. While the sensitivity for abusive head trauma of perimacular folds was 8% and of macular retinoschisis was 14%, their specificity was 100%, as the two findings were not seen in accidental trauma in any of the reviewed studies. However, these findings have been reported in case reports of fatal crush head injury, fatal motor vehicle accident with severe rotational and deceleration injury, and one fatal 11-meter fall, which also produced multiple skull fractures [17–20]. Therefore, when macular retinoschisis or retinal folds are present in a child not involved in a fatal car accident or who does not have multiple skull fractures (as from a high fall or crush injury), the positive predictive value of these findings for abusive head trauma remains 100% based upon the available literature. Finally, note that the absence of retinal hemorrhage does not rule out abusive head trauma.

Retinal hemorrhage generally cannot be accurately dated to determine the timing of the traumatic event. However, there are general patterns related to the resolution of retinal hemorrhage that can be helpful. Intraretinal hemorrhages in infants resolve very quickly, and even numerous hemorrhages can clear in a few days, while preretinal hemorrhages take much longer to clear and when predominant indicate chronicity of findings. Preretinal and vitreous hemorrhage can take weeks to months to clear.

Pathogenesis of retinal hemorrhage in abusive head trauma

It is unknown how much force it takes to create the ocular injuries seen in abusive head trauma. Different types of retinal hemorrhages may have different mechanisms and more than one mechanism may be involved in any given instance. The most likely contributing cause of retinal hemorrhages in abusive head trauma is vitreoretinal traction injury due to deceleration forces resulting from shaking and/or impact [3]. This theory is supported by the anatomical location of retinal hemorrhages, which are often concentrated in areas of strongest attachment between the retina and vitreous, including the macula, along the retinal vessels and at vitreous base in the far peripheral retina; the presence of macular retinoschisis and retinal folds, again centered in an area of strong attachment; and animal models of inertial injury demonstrating intraocular hemorrhage in areas of strong vitreoretinal attachment [3, 21].

Other proposed mechanisms include retinal venous obstruction from subacute or sudden rises in intracranial or intrathoracic pressure, direct spread of blood along the optic nerve and other factors such as vascular dysregulation. Venous occlusion is a classic cause of retinal hemorrhage in adults; however, central and branch retinal vein occlusion are rare in children and have characteristic appearances not consistent with many of the patterns of retinal hemorrhages seen in abusive head trauma. Increased intracranial pressure in children is associated with only superficial intraretinal hemorrhages located on or adjacent to swollen optic nerve head, a pattern that also does not match the widespread pattern of retinal hemorrhages seen in abusive head trauma [22]. A sudden rise in intracranial pressure, such as from aneurysmal rupture, may cause Terson syndrome, a term sometimes used to describe coincident intraocular and intracranial hemorrhage, though originally defined to be intraocular hemorrhage associated with a subarachnoid bleed. Terson syndrome is characterized by a preponderance of preretinal and vitreous hemorrhage, in a pattern that again does not match that of abusive head trauma, which consistently includes intraretinal hemorrhage when retinal hemorrhages are present. Tracking of blood from the intracranial space along the optic nerve sheath and into the eye is one hypothesized mechanism underlying Terson syndrome. In an autopsy study of 13 infants who died of abusive head trauma, optic nerve sheath hemorrhage was present only in the intraorbital portion of the nerve, a pattern inconsistent with continuous tracking of blood [2]. As mentioned above, in a review of 75 children with abusive head trauma, no correlation was found between the laterality or symmetry of retinal hemorrhage and intracranial hemorrhage [10]. A severe and abrupt increase in intrathoracic pressure may cause Purtshcer retinopathy. This mechanism has been proposed to be supported by rib fractures seen in some children with abusive head trauma. However, the primary finding in Purtscher retinopathy is white retinal patches, which are rarely if ever seen in abusive head trauma. In addition, many of victims of abusive head trauma do not have rib fractures.

Orbital shaking injury may play a role as well, possibly through disruption of autonomic nerves to the retinal vessels. The optic nerve is longer than the distance between the apex of the orbit and the back of the globe, allowing the eye and orbital contents to move when the child is shaken and cause injury from translational and rotational movements of the globe. Injury may preferentially occur at tethering locations, such as the optic nerve-globe junction and apex of the orbit. This mechanism might explain optic nerve sheath hemorrhage occurring predominately anteriorly along the nerve, which suggests that blood did not arise from communication within the intracranial space [23]. Optic nerve atrophy often seen in survivors of abusive head trauma may also be best explained by direct optic nerve injury within the orbit.

Differential diagnosis

Trauma is the most common cause of retinal hemorrhage in infants and young children. Aside from infants with birth-trauma related retinal hemorrhage, abusive head trauma is the most frequent cause, particularly if the hemorrhages are numerous, multilayered and extend to the retinal periphery. Other conditions are reviewed below.

Birth: Retinal hemorrhages are common in newborns. Overall, approximately one-third of newborns examined in the first two days of life have retinal hemorrhages; the proportion is higher in vacuum-assisted delivery (42.6%) and lower in vaginal deliveries (25.6%) [15]. Intraretinal hemorrhages begin to clear very quickly, so the proportion of neonates with retinal hemorrhage drops quickly with each week postpartum. Theories for pathophysiology include obstetrical and perinatal hemodynamic changes, ocular compression and decompression, and peripartum prostaglandin release. The retinal hemorrhage may initially be numerous and extend into the periphery, resembling abusive head trauma patterns. Preretinal and subretinal hemorrhages are relatively uncommon, and retinoschisis and folds are not seen as a result of birth. Intraretinal hemorrhages clear quickly, with 85% of eyes completely cleared within 2 weeks. Reports of intraretinal hemorrhages lasting more than three weeks involve only individual isolated intraretinal hemorrhages, with the rest having already cleared [24]. Therefore, numerous, diffuse intraretinal hemorrhages beyond 1 month of age are not due to birth and must be due to another cause. Preretinal and vitreous hemorrhage can take longer to clear.

Accidental head injury: Retinal hemorrhages may be seen with severe accidental injury. Multiple clinical and postmortem studies of eyes in patients with severe accidental head injury suggest that the rate of retinal hemorrhage is less than 4%, and in most studies the incidence is zero, particularly when the injury is due to a short fall [8, 9, 16, 25]. When retinal hemorrhages occur following accidental injury, the injury history is usually unambiguous and consistent (e.g., motor vehicle accident, witnessed very high fall), the clinical condition is life-threatening, other injuries match the traumatic forces, and the infants have no additional unexplained injuries. The retinal hemorrhages most often are confined to the posterior pole, few in number and rarely subretinal. More severe retinal hemorrhages, even with extension to the retinal periphery, have been reported with severe motor vehicle accidents involving multiple acceleration-deceleration events, such as vehicle rollovers [18].

Raised intracranial pressure: Elevated intracranial pressure causes only superficial intraretinal hemorrhages located on or adjacent to a swollen optic disc [22]. These retinal hemorrhages are due to whatever process is causing the raised intracranial pressure. Papilledema occurs in less than 10% of abusive head trauma cases. Retinal hemorrhage in the absence of disc swelling, and retinal hemorrhages that do not match this pattern, for example retinal hemorrhages located distant from the optic nerve, even elsewhere in the posterior pole, are not due solely to raised intracranial pressure.

Other systemic and ocular diseases: A number of systemic and ocular conditions may be associated with retinal hemorrhages but can be identified by other findings on ocular and physical examination, review of systems or laboratory testing. Examples include leukemia and other causes of coagulopathy, such as thrombocytopenia, severe anemia, clotting factor deficiencies and vitamin K deficiency. In general, retinal hemorrhages related to hematological abnormalities are less numerous, less extensive and do not extend far peripherally, with the exception of leukemia, which would easily be recognized by a routine blood count. Diabetic retinopathy is unheard of in infants. Hypertensive retinopathy features cotton-wool spots in the retina and is easily diagnosed on physical examination. Glutaric aciduria type 1 is a rare autosomal recessive metabolic disorder that is sometimes associated with subdural hemorrhage after minor head trauma. Pre-existing macrocephaly is a hallmark of the condition and is thought to place bridging veins on stretch thus making them more prone to shearing forces induced by mild head injury. Retinal hemorrhages may occur but are usually no more than a few pre- or intraretinal hemorrhages confined to the posterior pole. Although this disorder eventually results in serious neurological compromise, affected children may have normal development in early childhood, and early neurological signs are often subtle. Other examples include endocarditis, hemolytic uremic syndrome, meningitis, cerebral malaria, vasculitis, hemophagocytic lymphohistiocytosis and galactosemia. These conditions are rare in the abusive head trauma age range and can be eliminated as diagnoses through appropriate testing, which only need be pursued if the appropriate systemic symptoms and signs are present. The retinal hemorrhages in all these conditions are typically few and in the posterior pole. Ruptured vascular abnormalities that cause large amounts of intracranial hemorrhage, such as arteriovenous malformation and aneurysm, are rare in children and are associated primarily with preretinal and vitreous hemorrhage (see Terson syndrome above). Purtscher retinopathy may occur following severe acute compression injuries to the thorax, but while retinal hemorrhages may be present, white retinal patches are the predominant feature.

Cardiopulmonary resuscitation rarely produces retinal hemorrhages, even when it is prolonged and coagulopathy is present [26–28]. Resuscitation-related retinal hemorrhages are few in number and confined to the posterior pole [26]. Retinal hemorrhages following seizures are extremely rare, with an incidence across four studies of less than 0.1%, in which only one case had unilateral flame-shaped superficial retinal hemorrhages located just at the optic disc and nowhere else [29–32]. While valsalva retinopathy characterized by preretinal or vitreous hemorrhage (not intraretinal retinal hemorrhage) is a well-described entity in adults, excessive coughing has not been found to cause retinal hemorrhage in young children [33].

Outcomes

Children who have ophthalmological findings of abusive head trauma require close follow-up by an ophthalmologist. Dome-shaped retinal hemorrhages can break into the vitreous. Large hemorrhages in the vitreous can interfere with visual development and cause amblyopia in young children. Vitrectomy should be considered for these patients, particularly if the electrical response of the retina (electroretinogram) is good. The prognosis in such cases is guarded, even with vitrectomy. In general, visual potential in children with abusive head trauma can be limited by retinal scarring from retinoschisis and hemorrhage, optic nerve atrophy and cortical visual impairment, the latter considered to be the most frequent cause of visual impairment [3, 11, 13, 34]. However, the correction of refractive errors and treatment of amblyopia can help to maximize visual potential. While many children are considered severely visually impaired, the visual outcome is good in up to one-half of survivors.

Several eye findings seem to have prognostic neurological significance. The extent of intraocular hemorrhage, presence of macular retinoschisis lesions and presence of pupillary abnormalities at presentation have been correlated with fatal outcome and permanent neurological impairment [13]. The severity of retinal hemorrhages is associated with the severity of hypoxic-ischemic brain injury on diffusion-weighted MRI [22]. The correlation between severity of ocular injury and neurological outcome suggests a common traumatic mechanism of brain and ocular injury in abusive head trauma.

Retinal hemorrhage is an important diagnostic sign in infants. Ophthalmological consultation should be sought as soon as possible in children with intracranial hemorrhage or other signs suspicious for abusive head trauma, as retinal hemorrhages may resolve quickly. The presence of retinal hemorrhage beyond the neonatal period is highly suggestive of abusive head trauma, particularly as the severity of retinal hemorrhage increases. In general, the pattern of retinal hemorrhages carries valuable diagnostic information, which may help guide further systemic testing, imaging studies and forensic investigations, but it should be interpreted in the context of all the clinical history and findings through multidisciplinary child abuse team assessment.

Conflicts of interest None

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