

RETINAL FINDINGS IN ABUSIVE HEAD TRAUMA

Knowing what to look for could help to identify child abuse.

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Abusive head trauma (AHT), frequently termed shaken baby syndrome, refers to a constellation of clinical findings—classically the triad of subdural hemorrhage (Figure 1A), bilateral retinal hemorrhages, and anoxic encephalopathy—from which clinicians infer physical child abuse. Classic metaphyseal fractures of long bones

(Figure 1B) and rib fractures (Figure 1C), especially when they are in different stages of healing, and skin ecchymoses are other clinical findings suggestive of inflicted trauma.

Several cases of AHT were reported in the early 1970s by Guthkelch and Caffey, and, ever since, multiple iterations in nomenclature have been applied to reflect the evolution in our understanding of AHT and its mechanisms of injury.^{1,2} The initial terminology, whiplash shaken infant syndrome, evolved to shaken impact syndrome, then infant whiplash-shake syndrome, battered-child syndrome, and, even later, nonaccidental head trauma. Recently, AHT was coined to describe infants with the aforementioned clinical findings and other evidence of physical abuse. Although shaken baby syndrome is the most widely known term, AHT is preferred by the American Academy of Pediatrics, as it is thought to encompass all mechanisms of injury.³

A CHANCE TO MAKE A DIFFERENCE

The ophthalmologist's role is pivotal in assisting in the diagnosis of AHT. It is estimated that approximately 5% of child abuse cases present first to an ophthalmologist.⁴ The anterior segment examination is typically unrevealing in AHT. Anisocoria, poor pupillary reaction, and mydriasis often indicate severe concurrent central nervous system (CNS) depression.⁵ Documentation of funduscopy findings by an

ophthalmologist is critical in establishing a diagnosis in suspected cases of child abuse, given the medicolegal implications.

RETINAL FINDINGS IN AHT

The most common retinal manifestation of AHT is multiple retinal hemorrhages in multiple layers of the retina, seen in approximately 85% of cases. Sub-internal limiting membrane (sub-ILM), nerve fiber layer, intraretinal, and subretinal hemorrhages are seen frequently in different stages of healing, often with white centers (Figure 1D and E), which constitute fibrin aggregations.⁶ Retinal hemorrhages are frequently bilateral, although unilateral involvement does not rule out a diagnosis of AHT.⁷ Sub-ILM hemorrhages can break through into the posterior hyaloidal space or vitreous cavity, resulting in vitreous hemorrhage. Macular retinoschisis, often caused by intraretinal blood dissecting the outer nuclear layer from the inner retina, has been demonstrated with optical coherence tomography (OCT) and postmortem histopathologic analysis of victims of AHT.⁸ Retinal tears, retinal detachment, and (rarely) pigment epithelial detachments have been reported.^{9,10} Furthermore, the presence of circumferential retinal folds surrounding the macula has been regarded as a specific sign for AHT and has been associated with fatal outcomes.^{11,12} Optic disc hemorrhages and papilledema as a result of increased intracranial pressure may also be observed.

Recently, significant progress has been made in elucidating the underlying mechanisms leading to the retinal manifestations



AT A GLANCE

- Subdural hemorrhage, bilateral retinal hemorrhages, and anoxic encephalopathy, when seen together, are a triad indicating abusive head trauma (AHT).
- AHT is not the only condition associated with the presence of multilayered retinal hemorrhages.
- Ocular examination findings often provide compelling evidence in the diagnosis of AHT.

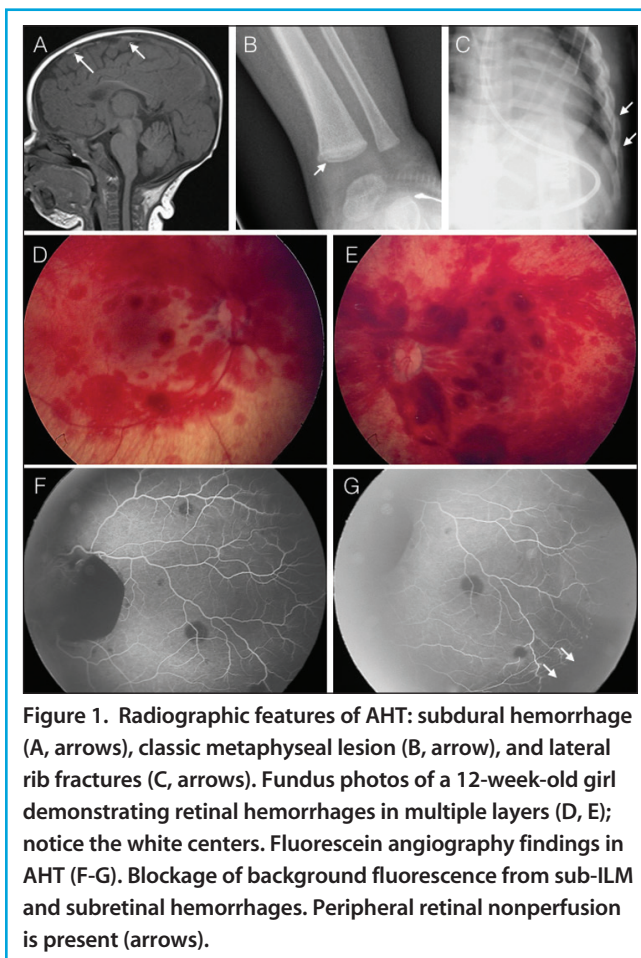


Figure 1. Radiographic features of AHT: subdural hemorrhage (A, arrows), classic metaphyseal lesion (B, arrow), and lateral rib fractures (C, arrows). Fundus photos of a 12-week-old girl demonstrating retinal hemorrhages in multiple layers (D, E); notice the white centers. Fluorescein angiography findings in AHT (F-G). Blockage of background fluorescence from sub-ILM and subretinal hemorrhages. Peripheral retinal nonperfusion is present (arrows).

of AHT. Animal and computer simulation models support the most widely accepted theory, that repeated accelerations and decelerations of the head and globe result in shearing forces between the retinal blood vessels and the tightly adherent infant vitreous.¹³ This hypothesis has been further supported by histopathologic and OCT findings of victims of AHT showing vitreoretinal membrane formation, vitreoretinal traction, and persistent attachment of the vitreous at the apices of perimacular folds.^{14,15}

Retinal neovascularization in response to peripheral retinal ischemia and nonperfusion has been identified as an important late clinical manifestation in infants with AHT, and this underlines the importance of evaluating these patients using widefield fluorescein angiography (Figure 1F and G).¹⁶⁻¹⁸ The mechanism behind the peripheral nonperfusion is not well understood. It is believed that several factors are involved, including venous stasis, direct vitreous shearing of the capillary network, and autonomic dysregulation of the retinal vascular network secondary to brain hypoxia and damage.¹⁸

DIFFERENTIAL DIAGNOSIS

The presence of multilayered retinal hemorrhages is not

pathognomonic for AHT, as several other conditions can present similarly. Terson syndrome (intraocular hemorrhage associated with intracranial hemorrhage) is one of the most important masqueraders of AHT, given the overlapping ophthalmic and CNS findings.¹⁹ Retinal folds have been reported in patients with Terson syndrome, but they are certainly much less common than in AHT.²⁰ Although the rare diagnosis of Terson syndrome has overlapping features with AHT, the literature and our experience both support the fact that AHT is more common and the likely diagnosis in almost all cases. Before a diagnosis of Terson syndrome can be made, a careful search (via clinical history, physical examination, skeletal survey, advanced CNS imaging, and consultations with a child abuse pediatrician and social services) for other findings of nonaccidental trauma must be made in order to avoid missing a potentially lethal diagnosis of AHT.

Multilayered retinal hemorrhages in infants can also be seen in the blood dyscrasias (eg leukemia and chronic anemias), vaginal delivery, sepsis, and blunt trauma. Therefore, an appropriate workup, along with careful history, should be performed to rule out these entities.²¹ Note that the pattern and extent of retinal hemorrhages are usually highly suggestive of AHT, especially when other retinal findings such as perimacular folds and retinoschisis are present along with the classic systemic signs.²²

Cardiopulmonary resuscitation (CPR) is occasionally performed on these often critically ill patients, so one question that commonly arises is whether CPR caused the retinal hemorrhages as a form of Valsalva retinopathy. Multiple studies have shown that retinal hemorrhages that develop after CPR are rare, and, when present, they are frequently confined to the posterior pole and are much less extensive than hemorrhages caused by AHT.^{23,24}

MANAGEMENT STRATEGIES IN AHT

When AHT is suspected, an ophthalmology consultation should be initiated early in the course of hospital admission in order to identify and document the retinal findings. Photodocumentation using a portable fundus camera is recommended. Depending on their extent, the majority of hemorrhages resolve spontaneously within a 1-to-3-week period; thus, observation only is usually recommended.

Persistent hemorrhage involving the fovea or dense vitreous hemorrhage may result in deprivation amblyopia, especially if involvement is unilateral. In such cases, surgical intervention with vitrectomy may be necessary to clear the hemorrhage if it does not clear on its own within 2 months.²⁵

If available, widefield fluorescein angiography is recommended to assess for peripheral ischemia. If nonperfusion is identified, then laser photocoagulation may be considered, depending on its extent, to avoid neovascularization and late sequelae.¹⁶⁻¹⁸

CONCLUSION

In nonfatal cases of AHT, long-term visual prognosis is

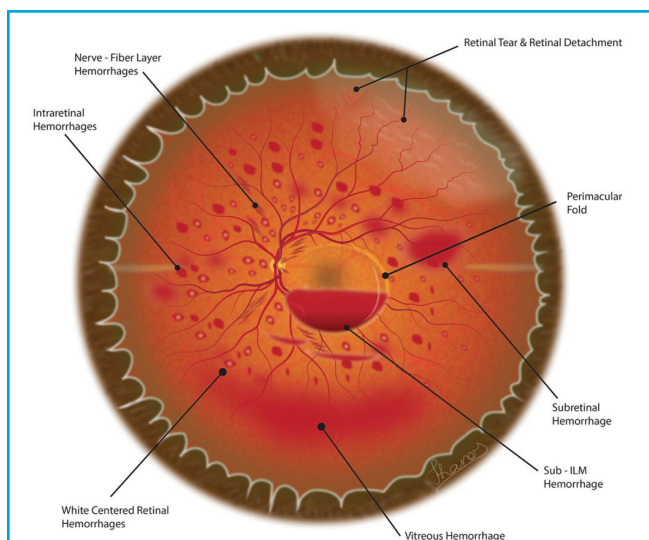


Figure 2. Illustration summarizing retinal findings seen in patients with AHT.

unfortunately often poor due to the high likelihood of concurrent traumatic brain injuries. Approximately 15% to 38% of victims succumb to their injuries, and, of the remaining patients, more than 80% will have permanent neurologic damage, possibly including blindness.^{26,27} Additionally, it has been shown that the severity and the extent of retinal hemorrhages correlates with the intracranial abnormalities of infants with AHT.²⁸

Despite recent advances in raising awareness and prevention, AHT continues to be a significant cause of preventable mortality and visual impairment in the pediatric population. AHT carries significant and often devastating implications that affect the patient's immediate family, relatives, and caregivers, and it requires the involvement of a multidisciplinary team of physicians including pediatricians, radiologists, neurologists, neurosurgeons, orthopedic surgeons, child protective services, and law enforcement agencies. The ophthalmologist's role is critical in confirming a diagnosis of AHT, as the dilated fundus examination findings (Figure 2) often provide compelling evidence. ■

1. Guthkelch AN. Infantile subdural haematoma and its relationship to whiplash injuries. *Br Med J*. 1971;2(5759):430-431.
2. Caffey J. On the theory and practice of shaking infants. Its potential residual effects of permanent brain damage and mental retardation. *Am J Dis Child*. 1972;124(2):161-169.
3. Christian CW, Block R; Committee on Child Abuse and Neglect; American Academy of Pediatrics. Abusive head trauma in infants. *Pediatrics*. 2009;123(5):1409-1411.
4. American Academy of Ophthalmology Quality of Care Secretariat, Hoskins Center for Quality Eye Care. Abusive head trauma/shaken baby syndrome - 2015. <http://www.aao.org/clinical-statement/abusive-head-traumashaken-baby-syndrome-june-2010>. Accessed February 9, 2016.
5. Kivlin JD, Simons KB, Lazaritz S, Ruttmann MS. Shaken baby syndrome. *Ophthalmology*. 2000;107(7):1246-1254.
6. Ling R, James B. White-centred retinal haemorrhages (Roth spots). *Postgrad Med J*. 1998;74(876):581-582.
7. Arloti SA, Forbes BJ, Dias MS, Bonsall DJ. Unilateral retinal hemorrhages in shaken baby syndrome. *J AAPOS*. 2007;11(2):175-178.
8. Koozekanani DD, Weinberg DV, Dubis AM, et al. Hemorrhagic retinopathy in shaken baby syndrome imaged with spectral domain optical coherence tomography [published online March 9, 2010]. *Ophthalmic Surg Lasers Imaging*.
9. Padrón Pérez N, Díaz-Cascasosa J, Prat-Bartomeu J, et al. Bilateral retinal detachment in a case of nonaccidental trauma. *Can J Ophthalmol*. 2013;48(3):e44-e45.
10. Ho LY, Goldenberg DT, Capone A. Retinal pigment epithelial tear in shaken baby syndrome. *Arch Ophthalmol*. 2009;127(11):1547-1548.
11. Levin AV. Retinal hemorrhage in abusive head trauma. *Pediatrics*. 2010;126(5):961-970.
12. Mills M. Funduscopic lesions associated with mortality in shaken baby syndrome. *J AAPOS*. 1998;2(2):67-71.
13. Nadarasa J, Deck C, Meyer F, et al. Update on injury mechanisms in abusive head trauma—shaken baby syndrome. *Pediatr Radiol*. 2014;44 Supp 4:s565-s570.

14. Emerson MV, Jakobs E, Green WR. Ocular autopsy and histopathologic features of child abuse. *Ophthalmology*. 2007;114(7):1384-1394.
15. Sturm V, Landau K, Menke MN. Optical coherence tomography findings in shaken baby syndrome. *Am J Ophthalmol*. 2008;146(3):363-368.
16. Brown SM, Shami M. Optic disc neovascularization following severe retinopathy due to shaken baby syndrome. *Arch Ophthalmol*. 1999;117(6):838-839.
17. Caputo G, de Haller R, Metge F, Dureau P. Ischemic retinopathy and neovascular proliferation secondary to shaken baby syndrome. *Retina*. 2008;28(3 Suppl):s42-s46.
18. Goldenberg DT, Wu D, Capone A, et al. Nonaccidental trauma and peripheral retinal neovascularization. *Ophthalmology*. 2010;117(3):561-566.
19. Weissgold DJ, Budenz DL, Hood I, Rorke LB. Ruptured vascular malformation masquerading as battered/shaken baby syndrome: a nearly tragic mistake. *Surv Ophthalmol*. 1995;39(6):509-512.
20. Keithahn MA, Bennett SR, Cameron D, Mieler WF. Retinal folds in Terson syndrome. *Ophthalmology*. 1993;100(8):1187-1190.
21. Agrawal S, Peters MJ, Adams GWW, Pierce CM. Prevalence of retinal hemorrhages in critically ill children. *Pediatrics*. 2012;129(6):e1388-e1396.
22. Levin AV, Christian CW; Committee on Child Abuse and Neglect, Section on Ophthalmology. The eye examination in the evaluation of child abuse. *Pediatrics*. 2010;126(2):376-380.
23. Odom A, Christ E, Kerr N, et al. Prevalence of retinal hemorrhages in pediatric patients after in-hospital cardiopulmonary resuscitation: a prospective study. *Pediatrics*. 1997;99(6):e3.
24. Longmuir SQ, McConnell L, Oral R, et al. Retinal hemorrhages in intubated pediatric intensive care patients. *J AAPOS*. 2014;18(2):129-133.
25. Capone A. Lens-sparing vitreous surgery for infantile amblyogenic vitreous hemorrhage. *Retina*. 2003;23(6):792-795.
26. Vitale A, Vicedomini D, Vega GR, et al. Shaken baby syndrome: pathogenetic mechanism, clinical features and preventive aspects. *Minerva Pediatr*. 2012;64(6):641-647.
27. Barlow KM, Thomason E, Johnson D, Minns RA. Late neurologic and cognitive sequelae of inflicted traumatic brain injury in infancy. *Pediatrics*. 2005;116(2):3174-e185.
28. Morad Y, Kim YM, Armstrong DC, et al. Correlation between retinal abnormalities and intracranial abnormalities in the shaken baby syndrome. *Am J Ophthalmol*. 2002;134(3):354-359.

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