Funduscopic Lesions Associated With Mortality in Shaken Baby Syndrome

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Purpose: The shaken baby syndrome (SBS) has been defined as a syndrome of intraocular and intracranial hemorrhage in young children, thought to be caused by violent shaking inflicted by an adult. In many cases SBS is fatal as a result of intracranial injury. Intraocular findings include hemorrhage, which may be accompanied by characteristic retinal folds or retinoschisis lesions. This study was performed to determine whether acute ophthalmologic findings might predict a fatal outcome. ***Methods:*** A consecutive series of 10 patients meeting a strict definition of SBS was reviewed for ophthalmic findings at presentation and outcome. ***Results:*** Seven patients survived, and three died. Of the six funduscopic characteristics identified in these patients, two were significantly associated with a fatal outcome: circular perimacular retinal folds found in four patients ($p = 0.048$) and peripheral retinoschisis lesions seen in three patients ($p = 0.012$). Lack of visual response at initial examination was also significantly associated with a fatal outcome ($p = 0.033$). ***Conclusions:*** Ophthalmic examination of children with suspected SBS is important for prognostic as well as diagnostic purposes. Circular perimacular retinal folds, peripheral retinoschisis lesions, and lack of visual response correlated with fatal neurologic trauma and may be useful in predicting severity of central nervous system injury in shaken baby syndrome. (J AAPOS 1998;2:67-71)

Shaken baby syndrome (SBS) is a term used to describe children younger than 3 years old with intraocular and intracranial hemorrhage without external evidence of direct blows to the eyes or head that cannot be explained by accidental trauma or other medical conditions.[1-4] The lack of external signs of trauma makes the ophthalmic examination of children with suspected child abuse crucial for the diagnosis of SBS.

Various patterns of intraocular hemorrhage are seen in SBS. Intraretinal, preretinal (subhyaloid), vitreous, and optic disc hemorrhage have been reported on funduscopy,[5-12] and combinations of these are commonly seen. Histopathologic examination has demonstrated intrascleral and subarachnoid hemorrhage.[5-7]

In addition to hemorrhage, three distinct retinal injuries have been described in nonaccidental trauma: retinal detachment,[13] circular perimacular retinal folds,[14] and retinoschisis.[15] Although there may be common features to the retinoschisis and circular perimacular retinal fold lesions, the distinctive appearance and consistent location of the perimacular folds allow separate ascertainment of these lesions. Other eye and vision abnormalities may include anisocoria, absent pupillary light response, and papilledema. None of the intraocular abnormalities are completely pathognomonic of SBS.[16-23] The diagnosis of SBS may be substantiated by a history supporting the mechanism of inflicted injury and forensic evidence of abuse.

Patients with acute SBS are frequently critically ill, comatose, or obtunded, with severe central nervous system (CNS) injury at the time of diagnosis. Mortality is frequent because of the intracranial axonal injury and complications of intracranial hemorrhage. Intracranial hemorrhage and CNS injury are usually evaluated with imaging studies, which may be difficult to perform or of suboptimal quality in the setting of the acute presentation. The extent of intracranial injury and the prognosis for survival and neurologic recovery are difficult to predict from the initial physical examination or early imaging studies.

The pattern of CNS injury in SBS is similar to that of the intraocular injury. A common mechanism, rapid acceleration and deceleration, causing shearing and traction on vitreoretinal attachments in the eyes and bridging meningeal vessels in the CNS, is thought to cause the hemorrhage seen in each.[13, 24] The common mech-

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TABLE 1. Age, ophthalmic findings, and outcome in SBS

Patient No	Age (mon)	No visual response	Pupil unresponsive	Pupils unequal	Vitreous hemorrhage	Preretinal hemorrhage	Retinal hemorrhage	Optic nerve hemorrhage	Circular retinal fold	Retinoschisis	Died
1	6	–	–	–	–	+	+	–	–	–	–
2	10	–	–	–	–	+	+	+	+	–	–
3	2	–	–	–	–	–	+	+	–	–	–
4	7	–	–	–	–	+	+	+	–	–	–
5	4	–	–	–	–	–	+	–	–	–	–
6	3	–	–	–	–	–	+	–	–	–	–
7	2	+	–	+	+	+	+				–
8	8	+	–	–	–	+	+	+	+	+	+
9	7	+	+	+	+	+	+	+	+	+	+
10	3	+	+	–	+	+	+	+	+	+	+

–, Finding not present; +, finding present in one or both eyes.

TABLE 2. Ophthalmic findings and statistical significance

Ophthalmic finding	No. of patients died with finding	No. of patients survived with finding	Statistical significance
No visual response	3	1	$p = 0.033$
No pupillary response	2	0	$p = 0.067$, NS
Anisocoria	1	1	$p = 0.300$, NS
Vitreous hemorrhage	2	1	$p = 0.183$, NS
Preretinal hemorrhage	3	4	$p = 0.475$, NS
Intraretinal hemorrhage	3	7	NS
Optic nerve hemorrhage	3	3	$p = 0.464$, NS
Circular perimacular retinal fold	3	1	$p = 0.048$
Peripheral retinoschisis	3	0	$p = 0.012$

p Values calculated with Fisher's exact test, two tailed. *NS*, Not significant, $p > 0.05$.

anism and response to injury would suggest that the severity of eye and CNS injury could be correlated in individual patients.

The extent of intraocular injury has been correlated with intracranial injury in patients who died of nonaccidental trauma.[13] In these patients the histopathologic intraocular abnormalities seen at autopsy, ranging from intraretinal and preretinal hemorrhage to vitreous hemorrhage and retinal detachment, were found to correlate with the extent and severity of intracranial injury.

By ranking the type and extent of fundus involvement, correlation between the severity of retinal hemorrhage and acute neurologic abnormalities has been demonstrated in one study.[25] Large preretinal hemorrhages, vitreous hemorrhage, and extensive fundus involvement were found retrospectively to be predictive of acute but not chronic neurologic involvement. However, this study did not incorporate retinal folds, retinoschisis, or other retinal lesions. In another small series dense vitreous hemorrhage was associated with poor visual outcome as a result of both intraocular and intracranial damage.[26]

This study was initiated to determine whether the extent of ophthalmic injury seen at the time of the initial eye examination might correlate with proximate mortality from acute SBS. The acute intraocular injury may identify infants with the greatest degree of injury and risk of neurologic morbidity and mortality. In addition, identification of intraocular and intracranial associations may allow insights into the mechanism of injury in SBS.

SUBJECTS AND METHODS

Ten consecutive patients seen at the University of Wisconsin Hospital from August 1993 to January 1997 with the diagnosis of traumatic retinal hemorrhage as a result of nonaccidental trauma were included in this study. All patients had undergone medial evaluation and had intracranial (subdural, subarachnoid, or intraparenchymal) hemorrhage on imaging studies (magnetic resonance imaging or computed tomography) as well as intraocular hemorrhage on ophthalmoscopy. Medical conditions causing bleeding were excluded by medical evaluation and systematic hematologic evaluation, when indicated. The history in each case was inconsistent with an etiology of accidental trauma. No patient had external evidence of significant direct head or eye trauma. Ophthalmic, radiologic, and neurologic evaluations were consistent with shaking as the mechanism of injury.[3, 4] Patients varied from age 2 to 10 months at the time of diagnosis, with a mean age of 5 months.

In addition to the medical evaluations, social work and police criminal investigations in each case concluded that the injury was not accidental. In 8 of the 10 cases criminal charges related to the injury were made.

I performed the ophthalmic examinations within 24 hours of hospital admission. Vision was tested with use of fixation lights, opticokinetic targets, and in cases of no response to other targets, a bright light (indirect ophthalmoscope on highest intensity). If there was no evidence of fixation, aversion, blink, or any other response to any visual stimulus in either eye, the patient was considered unresponsive. Pupils were tested with use of the indirect ophthalmoscope on high intensity in a dim room. Patients with no visible pupillary response in either eye were considered unresponsive. Asymmetric pupil size of 0.5 mm or greater was considered significant anisocoria.

Indirect ophthalmoscopy was performed after dilation of the pupils with tropicaimide 1% or a combination of tropicaimide 1% and phenylephrine 2.5%.

Ophthalmic findings and outcome of hospitalization (death during initial hospitalization vs survival to discharge) were analyzed retrospectively by review of examination notes and hospital records. Fisher's exact test was chosen to calculate p values to determine statistical significance, because of the small number of cases reviewed.

RESULTS

Complete records of eye examinations were available for each patient. However, dense vitreous hemorrhage in one patient (patient 7) precluded detailed examination of the macula and optic nerve. Details of the ophthalmic findings in each case are reported in Tables 1 and 2. Of the 10 patients included, three died of their acute CNS injuries 24 to 72 hours after their presentation.

All three patients who died and one survivor showed no visual response at the time of initial evaluation. The patient without response who survived (patient 7) has had persistently flat visual evoked potential, with occipital cortical atrophy on subsequent imaging studies. Lack of visual response was significantly associated with a fatal outcome ($p = 0.033$). Of the six patients demonstrating some visual response, four had fixation and following responses on lights or toys, and two responded with lid closure to bright light.

Two patients, both of whom died, had no pupillary response ($p = 0.67$, not significant [NS]). Two patients, one of whom survived, had initial anisocoria.

Indirect ophthalmoscopy was used to examine the extent of intraocular injury and hemorrhage. In one patient (patient 7), dense vitreous hemorrhage obscured the posterior pole in both eyes, and the patient could not be assessed for circular retinal folds, retinoschisis, or optic nerve hemorrhage.

The diagnosis of vitreous hemorrhage was made if there was sufficient hemorrhage within the vitreous space to be visible and interfere with the funduscopic view. One survivor and two patients who died had vitreous hemorrhage. Dense hemorrhage anterior to the retina but posterior to the posterior hyaloid without surrounding retinal elevation or whitening, seen over the macula or more peripheral retina, was considered preretinal hemorrhage and was seen in all fatalities and in four survivors. All patients seen in the study had hemorrhage within the neurosensory retina (intraretinal hemorrhage). Hemorrhages within the optic disc were seen in one eye of two patients and both eyes of another patient who survived and in all three fatalities. None of the forms of hemorrhage was found to be significantly associated with a fatal outcome (see Table 2).

No retinal detachments were seen in this series. Peripheral dome-shaped hemorrhagic lesions with white retinal borders (peripheral retinoschisis) and the circular perimacular retinal folds (Figure 1) were assessed separately. In all three patients with a fatal outcome, circular retinal folds were seen surrounding the macula in both eyes, with white margins and hemorrhagic centers. This finding was confirmed on autopsy examination of the eyes in each fatal case. The folds were continuous for at least 180 degrees, and in some cases completely circled the posterior pole of the retina. The apex of the folds appeared sharply elevated. These folds were consistent with the description of Gaynon et al.[14] This lesion was found to be significantly associated with a fatal outcome ($p = 0.048$). A similar appearance was seen in one eye of a patient who survived. Over the course of 6 months after the acute injury, the retina flattened with residual retinal pigment disturbance and loss of the foveal reflex. Best corrected recognition acuity in the affected eye is 20/40 after amblyopia therapy and 20/20 in the contralateral eye.

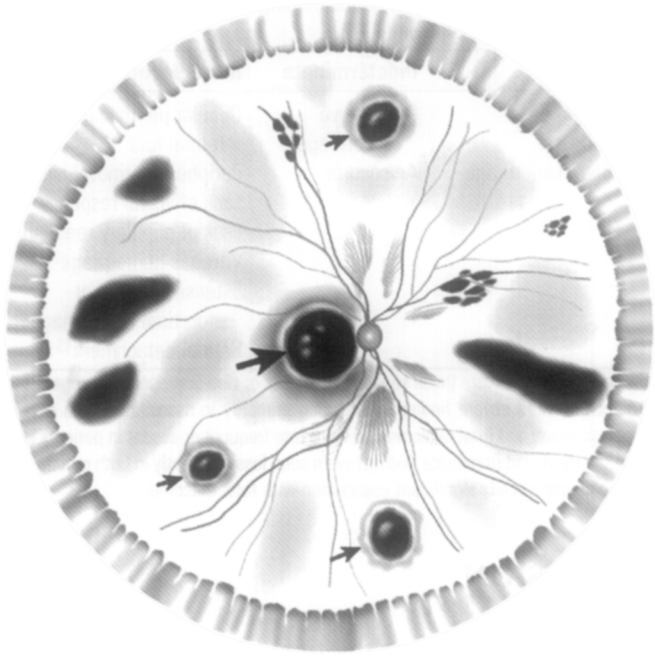

FIG. 1. Artist's rendering of retinal lesions seen in this series of patients with SBS. Drawing represents composite of abnormalities, including intraretinal and preretinal hemorrhage, circular perimacular fold *(large arrow)*, and peripheral retinoschisis lesion *(small arrows)*, drawn from clinical and autopsy photographs and drawings of patients included in this series.

Similar but smaller circular areas of dense hemorrhage surrounded by retinal elevation and whitening were seen in the retinal periphery in both eyes of all three patients who died and in none of the survivors ($p = 0.012$). These lesions were considered retinoschisis lesions, according to the criteria of Greenwald et al.[15] No survivor was found to have peripheral retinoschisis lesions on fundoscopy.

All three of the infants with fatal outcomes had the combination of retinal lesions (circular perimacular folds and peripheral retinoschisis) and absent visual response, and this combination did not occur in any of the patients who survived.

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TABLE 3. Classification of ophthalmic findings by severity of associated neurologic injury (fatal outcome)

Less severe	Indeterminate	More severe
Intraretinal hemorrhage	Optic nerve hemorrhage	Circular perimacular retinal fold
Preretinal hemorrhage	Anisocoria	Peripheral retinoschisis
		Lack of visual response
		Lack of pupillary reaction
		Vitreous hemorrhage[25, 26]
		Extensive retinal or preretinal hemorrhage[25]
		Retinal detachment[13]

Less severe findings were those seen in all patients or similar proportions, regardless of outcome. More severe findings were seen primarily in association with severe neurologic injury and fatal outcome and were less frequent or absent in patients surviving the injury. Indeterminate findings were seen less frequently in both groups and showed no statistically significant association with fatal outcome.

DISCUSSION

Funduscopic findings at the time of diagnosis may help determine the severity of neurologic injury and predict the likelihood of a fatal outcome from the neurologic injuries. In this retrospective series circular perimacular folds, peripheral retinoschisis lesions, and the lack of visual response on initial examination were associated with a fatal outcome from CNS trauma during the initial hospitalization.

None of the reported findings seen in SBS have been found to be completely pathognomonic, although the circular folds seen in this syndrome have been reported only under unusual circumstances without a nonaccidental injury.[23] Eye findings must be considered with the presenting circumstances and medical history and examination to exclude the possibility of trauma of accidental origin or medical condition simulating SBS. Forensic investigation should be confirmatory. In all cases included in this report, it was the conclusion of ophthalmic, pediatric, and forensic evaluation that the injury was inflicted by an adult. A mechanism of shaking was supported in all cases by the absence of external signs of blunt trauma.

Various mechanisms of intraocular hemorrhage and retinal lesions have been hypothesized in SBS. Increased intracranial pressure from the intracranial hemorrhage causing elevated retinal venous pressure, bleeding, and cystic dissection of blood within the retina has been postulated.[12, 27] However, severe retinal findings in patients without increased intracranial pressure (case 2) cannot be explained by this mechanism. The circular retinal folds are not always completely filled with blood, as would be expected with this proposed mechanism. Direct blows to the head in association with shaking have been thought to cause more severe intraocular damage.[17] None of the patients included in this series had skull fractures or external evidence of direct cranial trauma.

The role of acceleration/deceleration and traumatic vitreous separation on the development of circular retinal folds and retinoschisis has been supported by the observation of vitreous attachment to the apex of the circular perimacular folds in similar published cases of SBS[13, 24] as well as my observations of similar vitreous separation with persistent attachment and apparent traction on the folds in cases with autopsy histopathologic examination (unpublished data). Strong vitreoretinal adhesions exist in the perimacular and peripheral retina. These adhesions may be particularly tenacious in infants, who have a more formed vitreous than older children and adults.

Funduscopic findings significantly associated with a fatal outcome in this group includes peripheral retinoschisis and circular perimacular folds. These two abnormalities were seen in combination in three of four patients with the circular folds, and in all the fatalities. The similarity of these lesions and this frequent association would suggest that they may be identical in etiology, differing only in the site within the eye.

In all fatalities seen in this study the cause of death was neurologic injury from direct axonal trauma or intracranial hemorrhage. The close association of specific retinal injuries with death suggests that the severity of both ophthalmic and neurologic damage occurring in SBS may be directly related to specific factors. These factors may include the force of acceleration and deceleration of the head, rotational forces, anatomic features, and age. Mechanisms proposed for CNS injury in SBS are very similar to this hypothesis of intraocular injury. The movement of the brain within the cerebrospinal fluid during shaking injuries is restricted by the bridging blood vessels and overlying meninges. Inertial injury to the brain leads to bleeding when the vessels are stretched and neuronal injury from direct contusion or compression from the hemorrhage. Fatal injury, with the most severe intracranial injury and presumably the greatest force applied, is associated with a specific pattern of intraocular injury that is likely the result of a similar inertial mechanism.

In addition to funduscopic lesions, the lack of visual response is also associated with a fatal outcome. The lack of pupillary response, although not statistically significant, was more prevalent in patients with a fatal outcome. However, both these responses require intact and functional intracranial components of the visual pathway, which may be compromised by the intracranial injury independently from the ocular injury. These abnormalities are more likely to be directly related to the severity of the intracranial component of the injury than are the intraocular findings. In addition, the use of sedation or paralytic agents to treat these acutely ill patients who are frequently on artificial ventilation may interfere with assessment of pupils and vision.

This analysis also suggests an ophthalmic injury scale associating progressive eye findings with severity of neurologic injury, from less severe (intraretinal and preretinal hemorrhage) to more severe, associated with fatal neurologic injury (circular perimacular fold, peripheral retinoschisis, lack of visual response and absent pupillary reaction) (Table 3). In combination with the previously reported association of

severity of retinal hemorrhage,[25] vitreous hemorrhage,[26] and retinal detachment[13] with extent of neurologic injury, the presence of circular perimacular folds and peripheral retinoschisis should be considered significant indicators of the severity of SBS and markers of severe acute neurologic injury and risk of mortality.

The conclusions of this study are limited by the small number of patients studied. Further analysis of ophthalmic features and correlation with acute and chronic neurologic and CNS imaging findings may identify other associations of intraocular injury in addition to fatality. Development of an ophthalmic trauma score for SBS could allow more accurate prognostic estimation of acute and long-term CNS impairment in patients with SBS.

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NOTICE

After many years of valuable and dedicated service on the Editorial Committee of the AAPOS, the following individuals have been rotated off the Editorial Committee of AAPOS: Joseph H. Calhoun, MD; Jean Carruthers, MD; Eugene M. Helveston, MD; Keith McNeer, MD; Edward L. Raab, MD; and Arthur L. Rosenbaum, MD. The AAPOS is deeply appreciative of their hard work.

The Journal is pleased to announce that the following individuals have been added to the Editorial Committee: Edward G. Buckley, MD; James E. Egbert, MD; David A. Plager, MD; and Terri Young, MD. The Journal is also pleased to announce the addition of the following three distinguished strabismologists as international editors: Shinobu Awaya, MD, Kurashiki, Japan; Alberto O. Ciancia, MD, Buenos Aires, Argentina; and Joseph I. Lang, MD, Forch, Switzerland.