Prognostic Indicators for Vision and Mortality in Shaken Baby Syndrome

Craig F. McCabe, MD, PhD; Sean P. Donahue, MD, PhD

Objective: To study ocular and nonocular signs of patients diagnosed as having "shaken baby syndrome" and determine prognostic indicators for vision and mortality.

Methods: Medical records of child abuse cases involving bilateral retinal hemorrhages were reviewed. Particular attention was paid to visual function and pupillary light reaction at the time of admission as well as the location of retinal hemorrhages, neuroimaging findings, ventilatory requirement, and associated skeletal injuries. These findings were correlated with visual prognosis and mortality.

Results: Thirty consecutive cases met the criteria for review. At the initial visit, mean age of the children was 9.3 months (range, 1-39 months) and 12 children (40%) had at least fix-and-follow vision. Preretinal and intraretinal hemorrhages (93% [n = 28] and 100% [n = 30]) were more common than vitreous hemorrhage (10% [n = 3]). Subdural hematomas were detected in 21 patients (70%). Twenty children (67%) had seizures and 16 (53%) required ventilatory support; bruises and long bone fractures were seen in 14 (47%) and 4 (13%) children, respectively. Eight patients died. All patients with nonreactive pupils on presentation died, while all patients with a pupillary light reaction lived ($P<.001$). Six (86%) of 7 patients with midline shift died, whereas 21 (91%) of 23 with no midline shift lived ($P<.001$). At follow-up, retinal hemorrhages had resolved in nearly all children by 4 months, and 16 children (73%) had at least fix-and-follow vision. Ventilatory requirement was associated with poorer vision ($P<.01$).

Conclusions: Nonreactive pupils and midline shift of the brain structures correlate highly with mortality. Ventilatory requirement, but not visual acuity on presentation, predicts visual outcome.

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SHAKEN BABY syndrome (SBS) occurs following a severe shaking injury to children aged 3 years and younger and is seen with intracranial and retinal hemorrhages. This important form of nonaccidental trauma is difficult to diagnose because of its frequent lack of external signs.[1] Other associated findings may include diarrhea, bradycardia, hypothermia, hypotonia, irritability, seizures, bulging fontanels, and external and radiologic signs of physical abuse.[2,3] Histopathologic studies of postmortem eyes of children with SBS show ocular hemorrhages at the vitreous, preretinal, intraretinal, and subretinal layers as well as within the perineural sheath of the optic nerve and in the intrascleral perioptico region.[4-6] A comprehensive review of SBS was recently published.[7]

Several investigators have suggested that young children who are seen for apnea or coma and signs of trauma have a computed tomographic (CT) scan and dilated retinal examination performed by an ophthalmologist to rule out SBS.[8,9] Others have recommended that autopsies be performed on eyes from all small children who died without an obvious cause of death.[4] However, the current literature lacks large-scale studies that provide prognostic indicators for the immediate health and long-term visual potential of the abused child. In an effort to aid emergency department physicians, pediatricians, and ophthalmologists, we reviewed the charts of confirmed cases of SBS at Vanderbilt University Medical Center, Nashville, Tenn, during the past 5 years.

From the Department of Ophthalmology and Visual Sciences (Drs McCabe and Donahue), Pediatrics (Dr Donahue), and Neurology (Dr Donahue), Vanderbilt University School of Medicine, Nashville, Tenn.

RESULTS

Thirty patients (18 males and 12 females) with SBS met the criteria for our review. At presentation to our emergency department, their mean age was 9.3 months (range, 1-39 months). Twenty-two pa-

PATIENTS AND METHODS

We reviewed the medical records of 30 consecutive confirmed cases of SBS seen at the Vanderbilt University Medical Center between May 1992 and January 1998. These cases had bilateral retinal hemorrhages that had been photographed by the Department of Ophthalmology. A diagnosis of SBS was considered when bilateral retinal hemorrhages were observed in a situation where the injury was not consistent with the history, when other obvious signs of abuse were present, or if there had been a history of a previous suspicious episode in a lethargic infant. We routinely photograph the fundi of all children with retinal hemorrhages, so we believe no children were overlooked. The perpetrators of the abuse were determined by court records or by the social service and child abuse service notes in the hospital record. We evaluated the following characteristics at the initial visit: demographics (age, race, sex, and perpetrator's relationship), physical examination results (visual acuity, anisocoria, pupillary reactivity, ocular hemorrhage location, bruises, and hemiparesis), radiographic findings (intracranial hemorrhage location, presence of midline shift of the brain, skull fracture, and long bone fracture), and hospital course (ventilator requirement, seizure activity, and mortality).

Clinical follow-up occurred on a visit to the Vanderbilt pediatric ophthalmology clinic for 20 of the 22 living children, typically within 2 months of injury. The other 2 families were reached by telephone. At each follow-up visit until clinical findings became stable, the patient's vision, ocular motility, presence of amblyopia, and the resolution of intraocular hemorrhages were recorded for all survivors. The follow-up visits typically occurred when the children were quite young, which limited our ability to perform objective acuity testing in all patients and induced tropia testing in many others. Neuroimaging findings were determined by radiologists. Pupil examinations were performed by ophthalmologists, emergency medicine physicians, and pediatricians. Significant relationships between data variables were determined by χ^2 or Fisher exact tests.

Associated Findings in 30 Patients With Shaken Baby Syndrome

Associated Finding	No. (%) of Patients
Intraretinal hemorrhage	30 (100)
Preretinal hemorrhage	28 (93)
Subdural hemorrhage	21 (70)
Seizure	20 (67)
Ventilator requirement	16 (53)
Ecchymoses	14 (47)
Intracerebral hemorrhage	11 (37)
Subarachnoid hemorrhage	10 (33)
Skull fracture	8 (27)
Long bone fracture	4 (13)
Vitreous hemorrhage	3 (10)
Hemiparesis	1 (3)

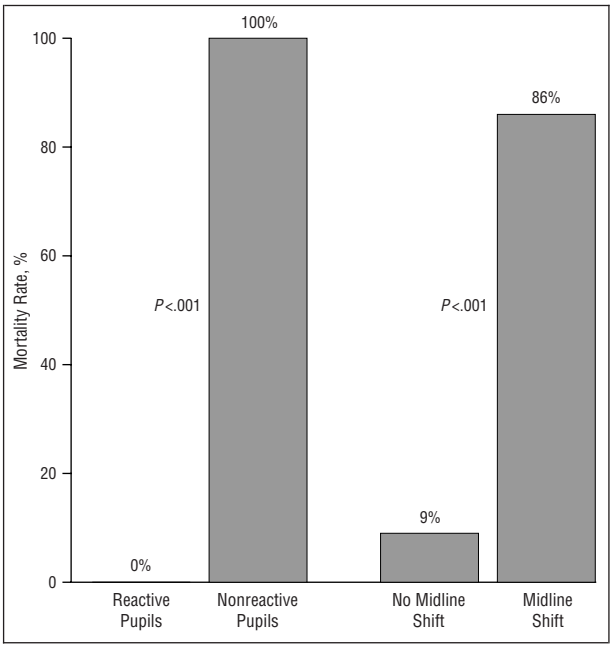

Figure 1. *Predictors of mortality in shaken baby syndrome. Both nonreactive pupils and midline shift of brain structures correlate with mortality.*

tients were white (73%), 7 were African American (23%), and 1 was Hispanic (4%). The race and sex distribution is similar to that of the Nashville population.

The perpetrator was identified in all 30 cases from social service notes and child abuse service notes placed in the chart at the time of discharge planning. The majority of these cases were successfully prosecuted, but individual case details are not available. Parents committed the child abuse in 23 cases (77%). The remainder of perpetrators included stepparents (n = 2; 7%), mother's boyfriend (n = 4; 14%), and babysitters (n = 1; 3%). In 18 cases (60%), the perpetrator was male. The Nashville population is 53% male.

The common findings associated with SBS are shown in the **Table**. All patients had intraretinal hemorrhages, but this was an inclusion criterion for this study. Preretinal hemorrhages were much more common than vitreous hemorrhages. Nonocular hemorrhages included subdural, intracerebral, and subarachnoid hemorrhages, as well as skin ecchymoses. During the children's hospital course, seizure activity and requirement for ventilatory assistance were common. Skull fractures occurred twice as often as long bone fractures.

Both pupillary nonreaction and midline shift of the brain at the time of presentation correlated highly with mortality (**Figure 1**). All 22 patients with reactive pupils survived, while all 8 patients with nonreactive pupils died. Most patients (21/23; 91%) without midline shift demonstrated by head CT survived, while most patients (6/7; 86%) with midline shift died. The 2 patients without midline shift who died had nonreactive pupils. Two of the patients who did not survive had midline shift of

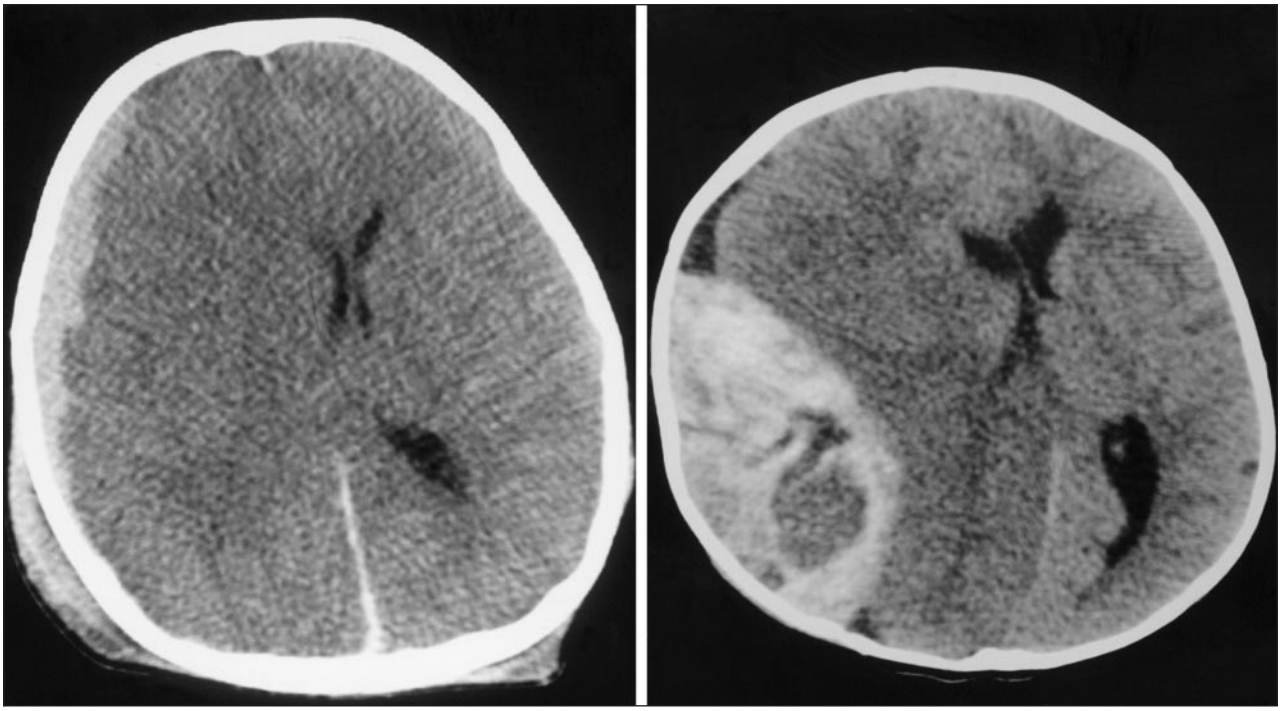

Figure 2. *Midline shift on neuroimaging in 2 children with shaken baby syndrome. Note the collapse of the occipital horn and diffuse intraparenchymal edema (left and right) as well as subdural hemorrhage and scalp hematoma (left). An acute epidural bleed (right) causes a flow void and midline shift. Both of these children died.*

the brainstem (**Figure 2**) and uncal herniation on initial neuroimaging.

Twenty of the 22 living children received follow-up examinations at Vanderbilt for at least 6 months. They were seen a mean ± SD 6.7 ± 8.2 months following the injury (range, 1-36 months). The remaining 2 were contacted by telephone. Retinal hemorrhages had resolved in 7 patients 1 month following injury, in 5 patients by 2 months following injury, in 4 by 3 months, 2 by 4 months, in 1 at the 9-month visit, and 1 patient 11 months following injury. (Many children did not have monthly examinations and this, therefore, represents the maximum time for hemorrhage resolution.)

In our population, 12 patients (40%) initially had fix-and-follow vision. Of those all lived, and 10 (83%) retained fix-and-follow vision, while 2 (17%) lost this vision in at least 1 eye after the retinal hemorrhages had resorbed. Of the 18 patients (60%) who presented without fix-and-follow vision in at least 1 eye, 8 died, and of the remaining 10 patients, 7 (70%) gained fix-and-follow vision and 3 (30%) never had improvement in their vision. Differences between these groups were not significant ($P>.6$). After resolution of retinal hemorrhages, approximately one fourth of the children (6 of 22) had poor vision in at least 1 eye. This was due to optic atrophy in 2 children, retinal fibrosis in 1, and traumatic cataract and retinal scarring in 1. Two patients had cortical visual impairment. No patients had macular folds. Interestingly, patients who did not require ventilatory support had better vision than those who required ventilation (**Figure 3**). At clinical follow-up, all 14 patients (100%) who did not require a ventilator had fix-and-follow vision, whereas only 4 (50%) of the 8 living patients who required ventilatory support during the hospital stay had good vision ($P<.01$).

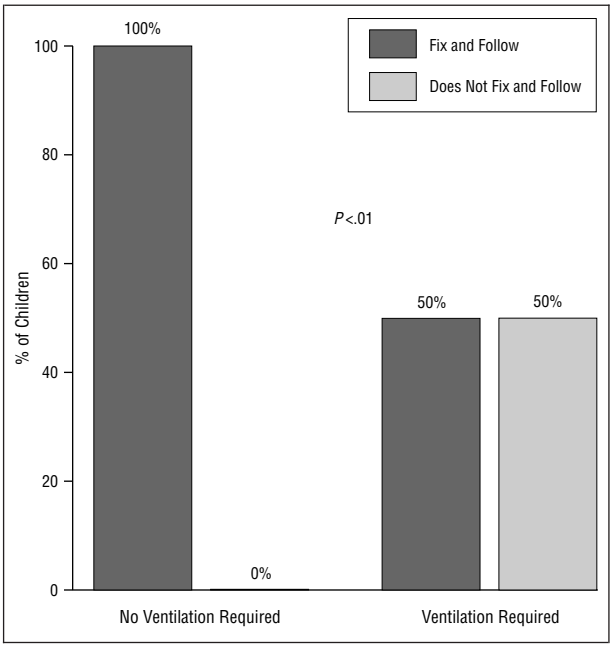

Figure 3. *Predictors of final vision in shaken baby syndrome. Lack of ventilator requirement is an indicator of good visual prognosis.*

COMMENT

This retrospective study describes the clinical and radiographic signs associated with SBS. In our review, 100% of patients with SBS with nonreactive pupils and 86% with

midline shift died. Thus, pupil reactivity and midline shift of brain structures are strong indicators of mortality in SBS. The mechanism for nonreactive pupils in severe SBS has not been described. Possibilities include bilateral afferent nerve trauma, dorsal midbrain lesions with light-near dissociation, uncal herniation, or generalized brain dysfunction. Since our initial CT scans did not consistently show chiasmal or midbrain abnormalities, and uncal herniation was detected in only 2 cases, the pathophysiological mechanisms of nonreactive pupils in SBS remain unclear.

Cases of SBS at our level I emergency department, located in a moderate-size city, mirrored the racial mix of the local population.[10] As reported by others,[11] we found males were the perpetrator in 60% of all SBS cases. In our study, only 47% of all patients with SBS had bruises and 13% had long bone fractures. Nonocular hemorrhages associated with SBS include subdural, subarachnoid, and intracerebral hemorrhages, and skin bruises. Our retinal findings agree with those previously reported that children with SBS have retinal hemorrhages at multiple levels.[8,12,13] The literature remains divided as to whether head trauma involving a direct blow by a hard object is required for SBS.[3,4,14]

Retinal hemorrhages resulting from accidental head trauma, seizures, cardiopulmonary resuscitation, and other etiologies must be ruled out when considering the diagnosis of SBS.[12,15-34] However, these retinal hemorrhages are typically quite different in appearance from those seen in SBS.[16,17,27,30,31] The history of present illness and extent of retinal hemorrhages typically narrow the differential diagnosis in SBS when the coagulation profile is normal. It is now widely believed that unexplained, extensive retinal hemorrhages in infants and young children are virtually diagnostic of nonaccidental trauma.[30-34]

Mills[35] and Matthews and Das[36] have recently reported prognostic indicators of vision and survival for shaken infants. Mills' series of 10 infants found specific retinal lesions associated with poor final vision and lack of visual response on presentation to be associated with mortality. Matthews and Das found that 3 of 5 patients with diffuse vitreous hemorrhages had light perception or no light perception vision and poor neurologic outcomes, but their small sample size prevented statistical evaluation. We found that all 8 patients who died were visually unresponsive at presentation; therefore, visual responsiveness in our series is highly suggestive of survival. This is likely because these children have less severe injuries to the brain. Cerebral injury and extensive preretinal macular hemorrhages have both been associated with profound vision loss in shaken infants.[13,37] Another contributor to poor vision is a macular fold, thought to arise from large preretinal hemorrhages after they have resorbed.[35,37] We did not see macular folds in any of our patients. Decreased vision in our patients was due to retinal scarring, optic atrophy, and cortical visual impairment. In our study, initial visual acuity did not predict final visual acuity; approximately three fourths of survivors eventually developed good vision independent of initial vision. However, because most children were too young to have Snellen acuity tested, it is possible they have subtle defects in acuity.

Shaken baby syndrome is a major cause of nonaccidental injury in children. It requires prompt recognition and management because of the risks of death and permanent neurologic and visual impairment. Our study suggests that any irritable, lethargic, or dyspneic infant seen in an acute care setting where SBS is a possibility should have a thorough pupil examination as part of the initial evaluation. If the pupils are nonreactive, ventilatory support should be readied and neurosurgery consulted as the child's life is likely in jeopardy. If the pupils are reactive, a head CT and ophthalmology consult for a fundus examination should be performed, although all children suspected of having SBS should probably have neuroimaging and an ophthalmology consult. Whether our protocol would influence outcome or even affect decision making in the acute care setting, however, is unknown, since other possible indicators of neurologic status would likely be present. In addition to our finding that survival is related to pupillary and radiographic signs, we also found that visual prognosis appears to be related to any requirement for ventilatory support. We hope this information will prove useful to emergency department physicians, pediatricians, and ophthalmologists responsible for the initial assessment and management of victims of child abuse.

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Corresponding author: Sean P. Donahue, MD, PhD, Department of Ophthalmology and Visual Sciences, 1215 21st Ave S, Nashville, TN 37232-8808.

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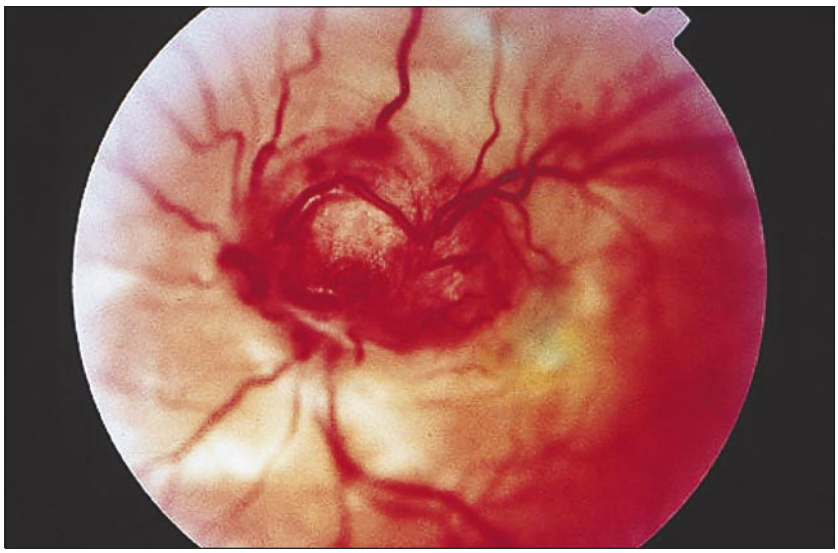
ARCHIVES Web Quiz Winner

Congratulations to our January Web quiz winner, P. Fritsche, MD, University Hospital Vrije Universiteit, Amsterdam, the Netherlands. The answer to the January quiz was capillary hemangioma and Von Hippel Lindau. For a complete discussion of this case, see the Case Reports and Small Case Series section in the February ARCHIVES (Malecha MA, Haik BG, Morris WR. Capillary hemangioma of the optic nerve head and juxtapapillary retina. *Arch Ophthalmol*. 2000;118:289-291).



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