

# Optic Nerve Sheath and Retinal Hemorrhages Associated With the Shaken Baby Syndrome

Scott R. Lambert, MD; Thomas E. Johnson, MD; Creig S. Hoyt, MD

• A 13-month-old child with the pathognomonic findings of the shaken baby syndrome died secondary to cerebellar herniation. Pathologic examination disclosed extensive intraocular, optic nerve sheath, and intracranial hemorrhages, despite a paucity of external signs of trauma. Many of the hemorrhages were old, suggesting that the child had experienced multiple episodes of trauma. Hemorrhages of the optic nerve sheaths have not been previously reported with the shaken baby syndrome but probably accompany this condition frequently. Our patient's ocular hemorrhages may have resulted from a sudden rise in intracranial pressure.

(Arch Ophthalmol 1986;104:1509-1512)

Child abuse is a common problem in our society and often has protean manifestations.<sup>1</sup> A frequently unrecognized form of child abuse occurs when infants are shaken; this has been referred to as the shaken baby syndrome.<sup>2,3</sup> Retinal hemorrhages are observed in a high percentage of these patients, along with subdural hematomas and cerebral edema, while there is often a paucity of external signs of trauma.<sup>4,5</sup> Retinal hemorrhages are also frequently seen in newborn infants but usually resolve within a few days without any serious sequelae.<sup>6,7</sup> In contradistinction, the retinal hemorrhages and ocular injuries that

occur following the shaking of infants may result in permanent visual handicaps due to the development of macular scarring,<sup>8</sup> retinal detachment,<sup>9</sup> and optic atrophy.<sup>10</sup> We report the clinical and pathological findings in an infant who had the pathognomonic findings of the shaken baby syndrome.

## REPORT OF A CASE

A 13-month-old girl was found to be unresponsive by her babysitter following an afternoon nap. The infant had vomited several times during the preceding four days but allegedly was "playful" earlier on the day of admission. A history of recent trauma or shaking was initially not elicited, but the babysitter later said that she had shaken the child vigorously four days before admission. The child's development had been normal.

On admission, the patient was noted to have tonic-clonic seizures, with limited spontaneous movements and a poor response to pain. Her only external signs of trauma were two ecchymoses on the mid-sternum. The right pupil was 6 mm in diameter and the left was 4 mm; both were nonreactive to light. No subconjunctival hemorrhages were present and the anterior segment was unremarkable. Indirect ophthalmoscopy demonstrated a large subhyaloid hemorrhage obscuring the right optic nerve and bilateral serosanguineous retinal detachments with extensive intraretinal hemorrhages.

Fig 1.—Computed tomographic scan depicting right hemispheric and left frontal lobe edema, midline shift to the left with collapse of right lateral ventricle, and subdural hematomas along falk cerebri (black arrow) and right hemisphere (white arrow).

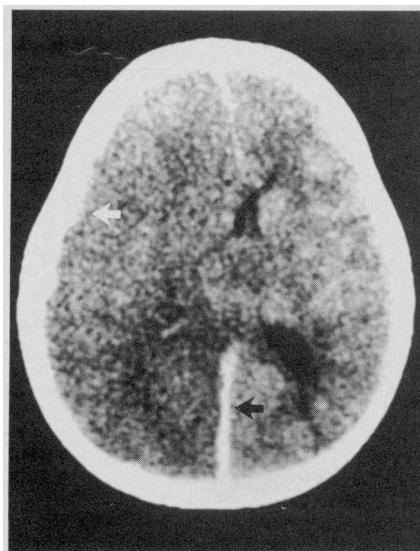


Fig 2.—Computed tomographic scan demonstrating intraocular hemorrhages in both eyes.



Accepted for publication May 22, 1986.

From the Department of Ophthalmology, University of California, San Francisco (Drs Lambert and Hoyt); and the Department of Ophthalmology, Pacific Medical Center, San Francisco (Dr Johnson).

Reprint requests to Room A704, University of California-San Francisco, 400 Parnassus Ave, San Francisco, CA 94143 (Dr Hoyt).



Figure 3.



Figure 4.

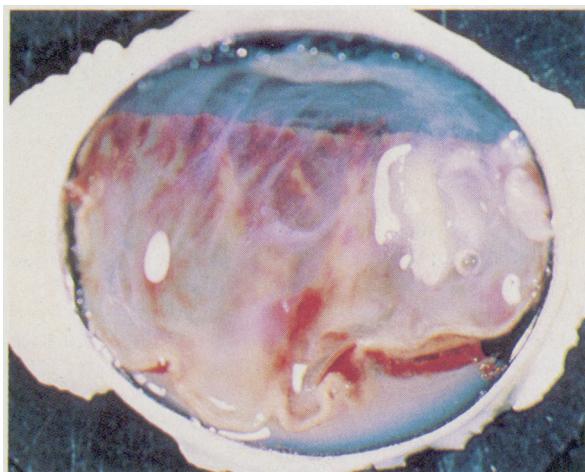


Figure 5.

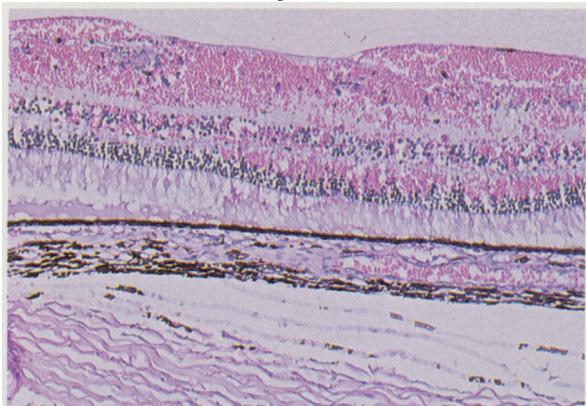


Figure 6.

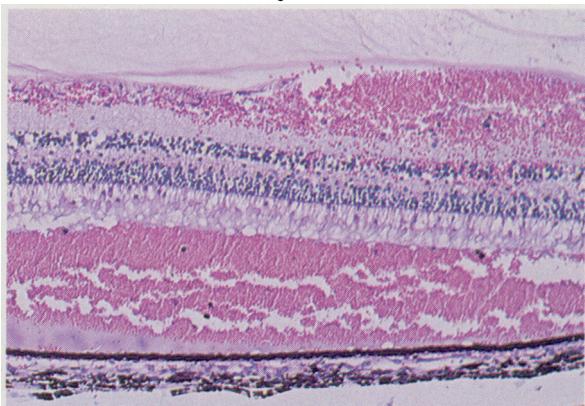


Figure 7.

Figure 8.

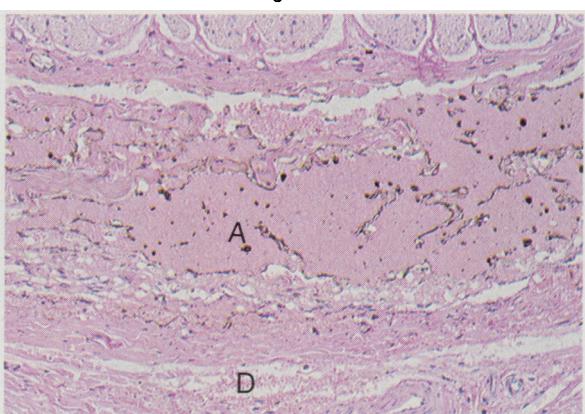
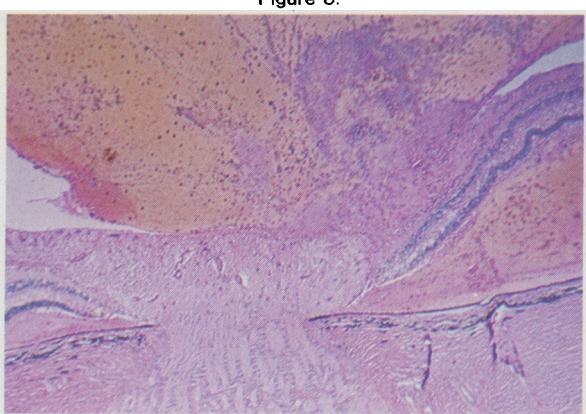


Figure 9.

Fig 3.—Intraorbital section of right optic nerve with old hemorrhage in perineural space.

Fig 4.—Left eye demonstrating retinal hemorrhages extending out to ora serrata and shallow serosanguineous detachments.

Fig 5.—Right eye with intraretinal hemorrhages, subhyaloid hemorrhage, and serosanguineous retinal detachment of posterior pole.

Fig 6.—Intraretinal hemorrhages primarily involving inner left retina (hematoxylin-eosin, X40).

Fig 7.—Shallow detachment of right retina by fresh hemorrhage (hematoxylin-eosin, X40).

Fig 8.—Papilledema of right optic nerve and old hemorrhages in both subretinal and prehyaloid space (hematoxylin-eosin, X25).

Fig 9.—Old and fresh hemorrhages in perineural subarachnoid space (A) and smaller amount of fresh hemorrhage in subdural space (D) (hematoxylin-eosin, X25).

A computed tomographic scan delineated a subdural hematoma along the right side of the calvarium and in the parieto-occipital interhemispheric space, edema of the right cerebral hemisphere and left frontal lobe, a marked midline shift to the left (Fig 1), and intraocular hemorrhages (Fig 2). There were no skull or long-bone fractures. Despite endotracheal intubation and the intravenous administration of mannitol and pressor agents, the patient died the following morning. Cardiopulmonary resuscitation was not administered. An autopsy was performed.

## RESULTS

Necropsy disclosed contusions overlying the sternum that were estimated to be three to five days old without associated thoracic or abdominal injuries. Gross examination of the brain showed a right subdural hematoma with a volume of 50 mL. The right cerebellar tonsil was softened and adherent to an edematous medulla oblongata, suggestive of herniation through the foramen magnum. Multiple focal subarachnoid hemorrhages were present over the right hemisphere near the midline. Microscopic examination of the subdural hematomas demonstrated erythrocytes lacking sharply defined borders, suggesting that they had been present for four to five days. In addition, widespread cerebral edema and focal collections of erythrocytes

in the cerebrum and subarachnoid space were noted.

The eyes were fixed in formaldehyde solution within 24 hours of the patient's death. Gross examination of the eyes before sectioning was unremarkable except for thickening of the perineural space with a brownish material resembling old hemorrhage (Fig 3). After horizontal sectioning, multiple areas of intraretinal hemorrhages extending to the ora serrata and shallow serosanguineous retinal detachments were noted (Figs 4 and 5). The vitreous was slightly hazy and there was a large preretinal hemorrhage obscuring the posterior pole in the right eye.

On microscopic examination the cornea, lens, and angle structures were unremarkable. Extensive areas of intraretinal hemorrhage involving primarily the inner retinal layers but in certain areas extending throughout the full thickness of the retina were observed (Fig 6). Multiple shallow retinal detachments were noted with underlying pools of intact and disintegrating erythrocytes (Figs 7 and 8) and an amorphous eosinophilic material. An old preretinal hemorrhage was also seen in the right posterior pole (Fig 8) with extension into the vitreous through the posterior hyaloid face. No cytid bodies were present and the choroid and sclera were normal. The subarachnoid space surrounding the optic nerve was filled with old and fresh hemorrhage and a small amount of fresh hemorrhage was present in the perineural subdural space (Fig 9). Both optic papillae were edematous (Fig 8), but there were no hemorrhages within the optic nerve parenchyma. No stainable iron was seen with a Prussian blue stain, suggesting that the hemorrhages were less than six days old.

## COMMENT

Shaking was first proposed as a cause of retinal hemorrhages in infants in 1967.<sup>2</sup> Subsequently, intracranial hemorrhages were shown to accompany these retinal hemorrhages frequently and the condition is now referred to as the shaken baby syndrome.<sup>11</sup> This syndrome most often occurs in children less than 18 months of age and is characterized by a dramatic disparity between external signs of trauma and the extent of central nervous system and ocular injuries.<sup>15</sup> While retinal hemorrhages are frequently seen for several days in newborns secondary to birth trauma,<sup>6,7</sup> they are uncommon in young children thereafter, except as a

sequela of child abuse.<sup>12</sup>

Our patient had the classic stigma of the shaken baby syndrome, including ecchymoses on the midsternum suggestive of thumbprints from being grasped around the chest, retinal hemorrhages, subdural hematomas, and cerebral edema. As is so common with this syndrome, her condition was not consistent with the history initially obtained because these injuries occur in "those too young to complain."<sup>13</sup> Ober<sup>14</sup> described a 10-month-old child presenting with lethargy and retinal hemorrhages, who, on pathological examination, had multiple retinal hemorrhages, hemorrhagic retinal detachments, an avulsion of the retina from the optic nerve, and a small subdural hematoma. Similarly, our patient had retinal hemorrhages dissecting through all layers of the retina and into the vitreous, serosanguineous retinal detachments, and subdural hematomas. Additionally, she had old and fresh hemorrhages in the perineural sheaths of the optic nerves, suggesting that she had experienced multiple episodes of trauma.

Several mechanisms have been proposed for the pathogenesis of retinal hemorrhages in the shaken baby syndrome. In 1967, Gilkes and Mann<sup>2</sup> postulated that they were caused by a sudden rise in intraocular venous pressure (such as in Purtzscher's retinopathy), either from shaking children while squeezing their chests or swinging them by their feet. In 1972, Caffey<sup>11</sup> noted the association between retinal hemorrhages and intracranial injuries in shaken infants, and later suggested that the head of an infant is particularly vulnerable to whiplash injuries<sup>3</sup> due to the proportionately larger head supported by poorly developed cervical muscles, the pliability of the sutures and fontanelles that allows for stretching of the calvarium, the greater deformity of the unmyelinated brain, and the increased percentage of cerebrospinal fluid. While all of these features of an infant's head allow greater shearing stresses to be exerted on the intracranial blood vessels, Caffey<sup>3,11</sup> did not elaborate on features of an infant's eye that might also predispose it to whiplash injuries. Others have shown that rapid deceleration alone can produce retinal hemorrhages<sup>15</sup> and Ober<sup>14</sup> proposed that the rapid acceleration and deceleration associated with the shaking of infants' heads, rather than an increase in venous pressure, may account for their hemorrhagic retinopathy.

Perlscher's retinopathy, occurring after crushing injuries to the chest, is characterized by cotton-wool spots and superficial hemorrhages.<sup>16</sup> Fluorescein angiography demonstrates diffuse capillary leakage and arteriolar staining several hours after onset<sup>17</sup> and impaired arteriolar flow, capillary nonperfusion, and venous staining several days later.<sup>18</sup> Burton<sup>18</sup> proposed that the retinopathy occurs secondary to arteriolar occlusion from air embolization, while others have attributed it to an acute rise in intraocular venous pressure, a secondary angiospasm, and subsequent hypoxia of the retina.<sup>16</sup> In contrast, Valsalva's hemorrhagic retinopathy, resulting from sustained closure of the glottis and a subsequent rise in intrathoracic pressure, is associated only with superficial retinal hemorrhages.<sup>19</sup> While Tomasi and Rosman<sup>20</sup> described two battered infants with retinal hemorrhages and "exudates," shaken infants typically have only retinal hemorrhages, suggesting a different etiology for the shaken baby retinopathy than for Perlscher's retinopathy. Likewise, the presence of hemorrhages extending throughout all layers of the retina and even into the subretinal space, in our patient and in the patient described by Ober,<sup>14</sup> compared with the usual superficial retinal hemorrhages associated with Valsalva's retinopathy, makes it unlikely that a Valsalva maneuver was solely responsible for these hemorrhagic retinopathies.

In 1958, Hollenhorst and Stein<sup>21</sup> reported the occurrence of retinal

hemorrhages in 51% of infants with intracranial hemorrhages, and Walsh and Hoyt<sup>22</sup> have emphasized that "when preretinal hemorrhages are found in an infant with or without cerebral symptoms, such a finding reliably indicates the presence of intracranial hemorrhage." Presumably, retinal hemorrhages occur after intracranial hemorrhages due to a sudden rise in intraocular venous pressure.

Optic nerve sheath hemorrhages are also commonly observed when the intracranial pressure rises suddenly; in fact, Muller and Deck<sup>23</sup> noted optic nerve sheath hemorrhages in 87% of eyes examined after a sudden rise in intracranial pressure, while only 37% of these eyes had intraocular hemorrhages. Walsh and Hedges<sup>24</sup> demonstrated that optic nerve sheath hemorrhages are rarely due to a direct extension of subarachnoid hemorrhage from the intracranial to intraorbital space; rather, they occur secondary to sudden distention of the subarachnoid space from an acute rise in intracranial pressure, which ruptures the dural and bridging vessels in the optic nerve vaginal sheaths. In contradistinction, intraocular hemorrhages occur only when the intracranial pressure rises still higher, resulting in occlusion of the central retinal vein and its chorioretinal anastomoses.<sup>25</sup> Optic nerve sheath hemorrhages probably occur more frequently with the shaken baby syndrome than has previously been recognized.

Our patient had a massive increase in her intracranial pressure, as

evinced by papilledema and cerebellar herniation, which undoubtedly contributed to the formation of her optic nerve sheath and retinal hemorrhages. Although an avulsed retina has been reported in a battered infant,<sup>16</sup> we did not note any ocular shearing injuries in our patient. In addition, even though ecchymoses on the patient's chest suggest that thoracic compression may have occurred during the shaking injury, cytoid bodies, which are classically seen with Perlscher's retinopathy, were not present. Finally, closure of the glottis from the child's crying while she was shaken may have contributed to the intraocular venous hypertension. While retinal hemorrhages may arise from multiple causes in the shaken baby syndrome, it seems likely that a sudden rise in intracranial pressure significantly contributed to their occurrence in our patient.

While only a small percentage of children with the shaken baby syndrome die of their injuries, many have residual neurological and ocular handicaps.<sup>26,27</sup> Optic atrophy, retinal detachment, and macular scarring may occur. Since retinal hemorrhages are often the only nonradiographic sign of shaking injuries in young children, ophthalmologists play an important role in the early recognition of the shaken baby syndrome.

We would like to thank Michael Drake, MD, Daniel Goodman, MD, William F. Hoyt, MD, and Alexander Irvine, MD, for their helpful comments in reviewing the manuscript.

## References

- Kempe GH, Silverman FN, Steele BF, et al: The battered-child syndrome. *JAMA* 1962;181:17-24.
- Gilkes MJ, Mann TP: Fundi of battered babies. *Lancet* 1967;2:468.
- Caffey J: The whiplash shaken infant syndrome: Manual shaking by the extremities with whiplash-induced intracranial and intraocular bleeding, linked with residual permanent brain damage and mental retardation. *Pediatrics* 1974;54:396-402.
- Zimmerman RA, Bilaniuk LT, Bruce D, et al: Computed tomography of craniocerebral injury in the abused child. *Radiology* 1979;130:687-691.
- Ludwig S: Shaken baby syndrome: A review of 20 cases. *Ann Emerg Med* 1984;13:104-107.
- Baum JD, Bulpitt CJ: Retinal and conjunctival hemorrhage in the newborn. *Arch Dis Child* 1970;45:344-349.
- Sezen F: Retinal hemorrhages in newborn infants. *Br J Ophthalmol* 1970;55:248-253.
- Harcourt B, Hopkins D: Permanent chorioretinal lesions in childhood of suspected traumatic origin. *Trans Ophthalmol Soc UK* 1973;93:199-205.
- Weidenthal DT, Levin DB: Retinal detachment in a battered infant. *Am J Ophthalmol* 1976;81:725-727.
- Harcourt B, Hopkins D: Ophthalmic manifestations of the battered-baby syndrome. *Br Med J* 1971;3:398-401.
- Caffey J: On the theory and practice of shaking infants: Its potential residual effects of permanent brain damage and mental retardation. *AJDC* 1972;124:161-169.
- Eisenbrey AB: Retinal hemorrhages in the battered child. *Child's Brain* 1979;5:40-44.
- Benstead JG: Shaking as a culpable cause of subdural hemorrhage in infants. *Med Sci Law* 1983;23:242-244.
- Ober RR: Hemorrhagic retinopathy in infancy: A clinicopathologic report. *J Pediatr Ophthalmol Strabismus* 1980;17:17-20.
- Lyle DJ, Stapp JP, Button RR: Ophthalmologic hydrostatic pressure syndrome. *Am J Ophthalmol* 1957;44:652-656.
- Marr WG, Marr EG: Some observations on Perlscher's disease: Traumatic retinal angiopathy. *Am J Ophthalmol* 1962;54:693-705.
- Kelley JS: Perlscher's retinopathy related to chest compression by safety belts. *Am J Ophthalmol* 1972;74:278-283.
- Burton TC: Unilateral Perlscher's retinopathy. *Ophthalmology* 1980;87:1096-1105.
- Duane TD: Valsalva hemorrhagic retinopathy. *Am J Ophthalmol* 1973;75:637-642.
- Tomasi LG, Rosman NP: Perlscher retinopathy in the battered child syndrome. *AJDC* 1975;129:1335-1337.
- Hollenhorst RW, Stein HA: Ocular signs and prognosis in subdural and subarachnoid bleeding in young children. *Arch Ophthalmol* 1958;60:187-192.
- Walsh FB, Hoyt WF: *Clinical Neuro-ophthalmology*. Baltimore, Williams & Wilkins Co, 1969, vol 3, p 2348.
- Muller PJ, Deck JHN: Intraocular and optic nerve sheath hemorrhage in cases of sudden intracranial hypertension. *J Neurosurg* 1974;41:160-166.
- Walsh FB, Hedges TR: Optic nerve sheath hemorrhage. *Am J Ophthalmol* 1951;34:509-527.
- Hayreh S: Pathogenesis of occlusion of the central retinal vessels. *Am J Ophthalmol* 1971;72:998-1011.
- Oliver JE: Microcephaly following baby battering and shaking. *Br Med J* 1975;2:262-264.
- Frank Y, Zimmerman R, Leeds NMD: Neuropathological manifestations in abused children who have been shaken. *Dev Med Child Neurol* 1985;27:312-316.