Vitreoretinal Traction and Perimacular Retinal Folds in the Eyes of Deliberately Traumatized Children

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Abstract: The pathophysiology of perimacular folds in eyes of deliberately traumatized children is disputed. The authors reviewed the clinical and forensic records and systemic and ocular findings at autopsy of three children with perimacular retinal folds who died after being violently shaken. Two of the children suffered direct head trauma in addition to being shaken; one patient was violently shaken without any physical or forensic evidence of direct head trauma. No direct ocular trauma was detected. In each case, the vitreous had partially separated from the retina but remained attached to the internal limiting membrane at the apices of the folds and the vitreous base, implicating traction in the pathogenesis of these folds. Although some intraocular findings in deliberately traumatized children may be explained by direct head injury, the possibility of both direct head trauma and shaking must be considered. Perimacular folds may develop without direct ocular or head trauma and may constitute evidence supporting violent shaking. *Ophthalmology 1991; 98:1124–1127*

Shaken baby syndrome is a form of child abuse characterized by minimal or absent signs of external injury, subdural hemorrhages with or without subarachnoid hemorrhages, and retinal hemorrhages, caused by severe shaking of infants by the extremities or shoulders.[1–5] In addition to hemorrhages located in the subdural space in the optic nerve, vitreous, and all layers of the retina, peculiar circinate perimacular folds have been described in cases of presumed shaken baby syndrome.[6] Clinically, these lesions were elevated, ring-shaped, white retinal ridges surrounding both maculas outside the vascular arcades. The children who developed these retinal folds suffered severe, indirect closed head trauma with intracranial hemorrhage and severe neurologic damage in the absence of significant signs of external trauma. The folds, considered to result from lateral displacement of the neurosensory retina by vitreous traction, were attributed to the shaking episode.[6]

In a recent study of eyes examined at autopsy, Elner et al[7] suggested that intraretinal hemorrhages, retinoschisis, and traction retinal folds may be the result of direct trauma to the skull and that the extreme acceleration–deceleration forces necessary to produce these changes are probably not solely generated by shaking. The medicolegal implications of this supposition are significant because the determination of innocence or guilt of an accused caretaker may depend on knowing whether a child was shaken. To determine the validity of the conclusions drawn by Elner et al[7] in their study, we correlated the ocular pathology findings in three patients who had macular folds and who

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died after episodes of severe shaking with systemic autopsy findings and the results of forensic investigations.

CASE REPORTS

Case 1. The caretaker of a 7.5-month-old boy claimed that the child suddenly "went limp." Cardiopulmonary resuscitation was initiated, and the child was intubated before being transferred to The University of Iowa Hospitals and Clinics because of seizures, unresponsiveness, and lack of respiratory effort. After arrival at the hospital, the child displayed opisthotonic posturing, inconsistent and transient extensor posturing in response to pain, fixed and dilated pupils, and bilateral retinal hemorrhages. A computed tomographic scan of the head was consistent with bilateral subdural effusions and acute subarachnoid hemorrhage. A magnetic resonance image confirmed central herniation and bilateral subdural hematomas. He was subsequently declared brain dead, and supportive measures were discontinued.

Results of postmortem examination performed 1.5 hours after death showed intracranial subdural hematomas, diffuse subarachnoid hemorrhages, cerebral edema, and bilateral tonsillar and uncal herniation. No skull fractures were identified and there was no evidence of subgaleal hemorrhage. One eye was opened in the standard pupil–optic nerve plane, and the other eye was opened by an alternative technique originally designed to simulate the funduscopic view of posterior pole melanomas.[8] Both section planes showed retinal hemorrhages over the optic nerve and retinal folds (Fig 1). There was no gross or microscopic evidence of anterior segment trauma. Subdural and subarachnoid hemorrhages were detected in the optic nerve. Results of microscopic examination showed papilledema, focal serous retinal detachments in the posterior pole, scattered intraretinal and vitreous hemorrhages, and perimacular retinal folds. Vitreous was separated focally from the internal limiting membrane of the retina but remained adherent to the apex of the fold; a hemorrhagic retinoschisis cavity was identified beneath the internal limiting membrane (Fig 2).

A complete forensic investigation determined that the child had been violently shaken.

Case 2. A 9-month-old girl was in the care of a 12-year-old babysitter who stated that the infant rolled off the couch and landed on her milk bottle. When further questioned by the infant's mother, the babysitter stated that the infant may have slipped in the bathtub and hit her head. Still later, the caretaker mentioned shaking the infant and throwing her to the floor.

When paramedics arrived, the infant was apneic, bradycardic, and pale. By the time the infant reached the emergency room, she was comatose and was intubated. Several ecchymoses were identified on her left cheek and chest. Her pupils were sluggishly reactive to light, and bilateral retinal hemorrhages and perimacular retinal folds were noted (Fig 3). Skull x-rays showed multiple occipital fractures, and a computed tomographic scan of the head showed prominent subdural falcine blood. The patient's pupils became middilated, fixed, and nonreactive to light over the course of the next 24 hours. Electrocerebral silence was documented by two electroencephalograms, taken 24 hours apart. The patient died, and a postmortem examination was conducted 15 hours after death.

Bilateral parietal and occipital bone fractures were identified together with parietal subgaleal, subperiosteal, subdural and subarachnoid hemorrhages. The brain was edematous and dural sinus thrombosis was identified. Gross examination of the body showed nine paraspinal ecchymoses (Fig 4) overlying fractures of the right seventh, eighth, and ninth ribs (Fig 5).

External examination of the eyes showed bilateral subdural optic nerve hemorrhages; both eyes were opened by the alternative gross examination technique.[8] The interior of both eyes was remarkable for perimacular retinal folds (Fig 6), and intraretinal hemorrhages just posterior to the ora serrata (Fig 7). The vitreous was partially separated from the internal limiting membrane, but remained so adherent to this structure at the apex of the fold that the membrane was torn away from the retina (Fig 8). The vitreous that was attached to the free end of the internal limiting membrane over the fold could be traced anteriorly to the retina just posterior to the ora, the site of subinternal limiting membrane hemorrhages identified on the gross examination.

Case 3. A 13-month-old apneic and unresponsive girl was brought to the emergency room after a babysitter claimed to have found the infant on the floor. Results of physical examination showed bilateral skin contusions around the ears and bilateral retinal hemorrhages. Subdural and subarachnoid hemorrhages were noted on computed tomographic examination. The child died within 29 hours of admission.

Examination of the body at autopsy, performed 12 hours after death, disclosed contusions on both ears at the helices, on both cheeks, and on the anterior axillary folds and on the right triceps. A subgaleal hemorrhage was noted over the occiput and in the left temporal scalp near the ear. Bilateral subdural and subarachnoid hemorrhages accompanied cerebral edema; there was no evidence of skull fracture. Both eyes were opened by the alternative examination technique[8] disclosing bilateral circinate perimacular retinal folds and peripheral retinal hemorrhages, identical to those previously observed (cases 1 and 2). Results of microscopic examination showed blood in all retinal layers, in the subhyaloid region, and in the vitreous in the retinal periphery. The vitreous, partially separated from the retina, remained adherent to the internal limiting membrane at the apex of the retinal folds and could be traced anteriorly to the subinternal limiting membrane hemorrhage in the retina just posterior to the ora serrata. Blood was detected in the subdural and subarachnoid spaces of the optic nerve. The anterior segment was unremarkable.

Although the child had no skull fractures, the presence of subgaleal hemorrhages and contusions on the cheeks and around the ears indicated direct trauma to the head. The contusions on the anterior axillary folds and around the triceps indicated that the child had been grasped in these areas. Contusions around these regions of the extremities are considered evidence of violent shaking.[1–5]

DISCUSSION

Retinal folds may appear as a postmortem artifact. Such folds appear to originate at the optic nerve head and radiate like spokes from the hub of a wheel. By contrast, the folds characteristic of violent shaking typically arch over the macula as illustrated in Figures 1, 3, and 6.

Our study confirms the role of vitreous traction in formation of perimacular folds,[6,7] as evidenced by the partial vitreous detachment except at the apices of the folds (Figs 2, 3). Additionally, the massive retinal hemorrhage at the vitreous base (case 2, Fig 7), a finding not previously described in eyes of deliberately traumatized children, also may be explained by vitreous traction: a strand of vitreous connected the hemorrhage at the ora to the apex of the folds. The vitreoretinal attachments between the posterior hyaloid face and the internal limiting membrane are particularly tenacious in infant eyes[9] and account for the posterior location of the perimacular folds as well as the

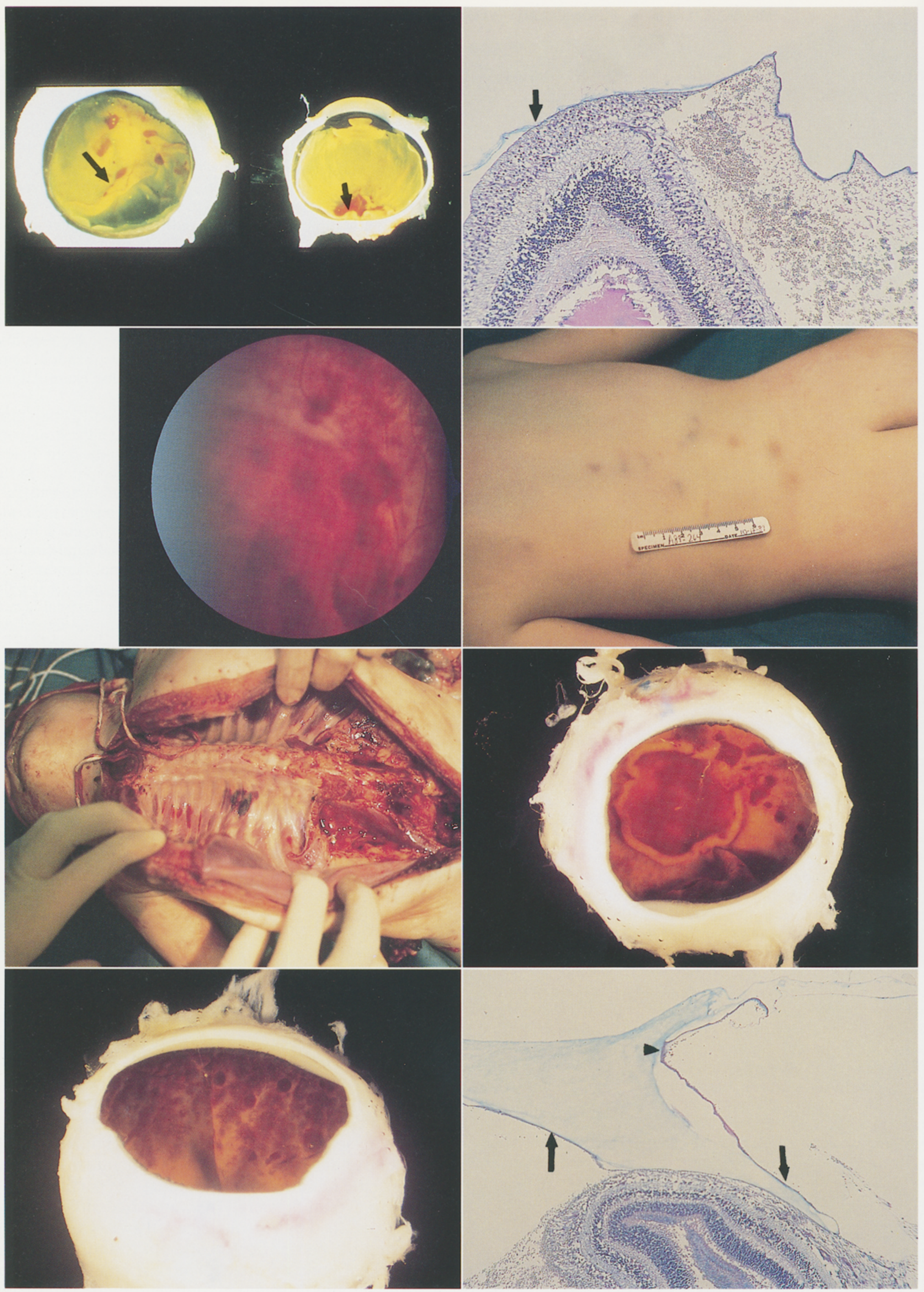
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Fig 1. *Top left,* gross ocular findings in both eyes of a child (case 1) who was violently shaken but who sustained no direct trauma to the eyes or head. Notice the perimacular folds (arrows) and hemorrhages (courtesy of The University of Iowa Ophthalmology Videodisc Project II, University of Iowa, 1990). **Fig 2.** *Top right,* histologic section through the perimacular fold in the right eye (case 1). The vitreous is partially separated from the internal limiting membrane (to left of arrow), and the internal limiting membrane is separated from the rest of the retina by a hemorrhage (colloidal iron-periodic acid Schiff; original magnification, ×100). **Fig 3.** *Second row left,* clinical fundus photograph of the right eye from case 2 taken on admission to The University of Iowa Hospitals and Clinics with a hand-held fundus camera. Notice the retinal hemorrhages and perimacular retinal fold. **Fig 4.** *Second row right,* back of a child (case 2) demonstrating paravertebral ecchymoses. The child also sustained paraspinal rib fractures (Figure 5). **Fig 5.** *Third row left,* paravertebral rib fractures, sustained as a consequence of violent shaking (case 2). **Fig 6.** *Third row right,* gross examination of the right eye from case 2. Notice the hemorrhages and the circinate macular fold (courtesy of The University of Iowa Ophthalmology Videodisc Project II, University of Iowa, 1990). **Fig 7.** *Bottom left,* same eye as illustrated in Figure 5. Notice the intraretinal hemorrhages just posterior to the ora serrata (courtesy of The University of Iowa Ophthalmology Videodisc Project II, University of Iowa, 1990). **Fig 8.** *Bottom right,* histologic section through the perimacular fold in the left eye (case 2). The internal limiting membrane has been stripped from the inner surface of the retina (arrows). The vitreous remains focally attached to the internal limiting membrane (arrowhead; colloidal iron–periodic acid Schiff; original magnification, ×63).

formation of subinternal limiting membrane hemorrhages at the vitreous base.

The pathogenesis of perimacular folds in the eyes of children subjected to deliberate trauma is disputed. Based on clinical observations only, Gaynon et al[6] concluded that shaking alone could generate enough vitreous traction to produce these folds. Elner et al[7] concluded that "some ocular injuries believed to result from shaking alone may, in fact, be due to direct head injury."

The infant described in case 2 was clearly shaken, as evidenced by the distinctive paraspinal ecchymoses and paraspinal posterior rib fractures, findings that implicate anterior–posterior trunk compression by fingers. This infant also was deliberately struck in the head, as evidenced by the skull fractures. This child, therefore, suffered a combination of shaking and direct head trauma. The pattern of cutaneous contusions discovered at autopsy in case 3 indicates direct blows to the head as well as grasping around the shoulders and extremities, a maneuver frequently seen in children who are violently shaken. This infant, like the one reported in case 2, also suffered a combination of direct trauma and shaking. It is well known that direct head trauma is frequently accompanied by shaking.[10–12] However, the formation of perimacular folds in both eyes of an infant who was violently shaken, but whose eyes and skull were not directly traumatized, proves that shaking alone is sufficient to produce these folds.

Because perimacular folds may develop after shaking alone, perimacular folds may be diagnostic of shaking. The best way to confirm this assertion is to study a series of eyes taken from children who sustained accidental (not deliberate) direct head trauma: if perimacular folds are not seen in this group, then it is reasonable to conclude that direct head trauma alone without shaking is insufficient trauma to induce the type of vitreous traction necessary to produce these folds.

The relationship between shaking and perimacular folds is not merely academic. A common legal defense for caretakers accused of abuse is the assertion that an object fell on the child or that the child fell against a hard surface (e.g., slipped in the tub). With subgaleal hemorrhages and skull fractures detected clinically or at autopsy, it may be difficult for prosecuting attorneys to prove deliberate trauma. The finding of perimacular folds, however, would suggest deliberate rather than accidental trauma if a large autopsy series can demonstrate that these folds do not appear in children who are victims of accidental direct head trauma. Shaking alone, proven by us to cause these folds, is never an accidental phenomenon. The purpose of distinguishing between accidental and deliberate trauma is important because a falsely accused caretaker may suffer the consequences of a conviction, while a guilty caretaker may be exonerated unless deliberate trauma (i.e., shaking) can be proven. Because many abusers repeat their actions, the false acquittal of a caretaker who has deliberately abused a child may subject other children to injury.

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