# Postmortem Orbital Findings in Shaken Baby Syndrome

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• PURPOSE: To compare postmortem orbital findings in pediatric accidental head injury to Shaken Baby Syndrome (SBS).
• DESIGN: Retrospective study.
• METHODS: SETTING: Institutional. STUDY POPULATION: Thirty-six patients underwent postmortem modified exenteration with sectioning of the orbital contents; 18 victims of SBS and 18 cases of fatal accidental head trauma. OBSERVATION PROCEDURE: In all cases of children who died from accidental head trauma, the orbital tissues were separated to expose the optic nerve sheath. Patients with gross evidence of hemorrhage within the sheath were included. All cases of SBS were included. After accidental head injury, exenteration was performed only if optic nerve sheath hemorrhage was suspected on gross examination. All children younger than 18 years old with head injury as primary cause of death were included. SBS is defined as having at least two of the following: (1) typical abnormal findings on neuroimaging, (2) typical skeletal injury, (3) retinal hemorrhages, (4) history of abusive shaking with or without blunt head trauma, or (5) an inadequate history to explain the observed injuries. MAIN OUTCOME MEASURE: Presence or absence of orbital hemorrhage.
• RESULTS: Orbital tissue injury is more common in SBS than accidental head trauma without orbital fracture. In addition, optic nerve sheath and optic nerve intradural hemorrhage are also significantly more common in SBS ($P < .0001$).
• CONCLUSIONS: Our study reports new evidence of injury to orbital tissues in SBS and supports the concept that these finding are due to unique acceleration-deceleration forces of this type of abusive head injury. (Am J Ophthalmol 2006;142:233–240. © 2006 by Elsevier Inc. All rights reserved.)

RETINAL HEMORRHAGES ARE ONE OF THE PRINCIPLE manifestations of Shaken Baby Syndrome (SBS), occurring in 50% to 100% of cases, with the higher incidence figures being reported for more severely injured or deceased children.[1–5] Other ocular findings include schisis of the macula, optic atrophy, and, less commonly, retinal detachment. Although the precise pathologic mechanisms are not fully understood, retinal hemorrhages in SBS are thought to be at least in part the result of acceleration-deceleration forces. For example, in traumatic retinoschisis, the uniquely firm pediatric vitreous-retinal adherence leads to splitting of the retina layers, most often at the level of the internal limiting membrane. Abusive blunt head impact may amplify the magnitude of the acceleration-deceleration forces.[6] However, impact is not required to induce the injuries that characterize SBS.[7,8]

In most reported pathology studies, the globe was removed via an anterior approach, after a conjunctival peritomy, with a limited portion of anterior optic nerve. When an intracranial approach has been used, the globe and a longer segment of optic nerve are usually removed and dissected away from the orbital contents. Trauma to the remaining orbital contents would go largely undetected. Because the orbit contains vital vascular and neurologic structures involved with retinal blood supply and autoregulation, and because the human globe can easily be translated in the orbit and is attached to an optic nerve with a length greater than the distance to the orbital apex, we hypothesized that violent shaking of an infant could result in injurious acceleration-deceleration forces. Movement to the globe resulting in damage to orbital tissues

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could have important implications for understanding the pathogenesis of retinal hemorrhage.

Although one author[9] has previously briefly reported in a book chapter a technique to open the globe from an intracranial approach through the orbital roof, we present a novel method for removing the intact orbital tissues and globe *en bloc* via a combined anterior and intracranial approach. We compared the findings in cases of fatal pediatric accidental traumatic head injury with SBS. Our study reports new evidence of possible shaking-related injury to orbital tissues.

## METHODS

ALL FORENSIC POSTMORTEM EXAMINATIONS FROM 1989 TO 2000 were undertaken by a protocol approved by the Ontario Coroners office, and performed by a single staff pathologist (C.S.) who was the designated forensic pathologist for these cases at our institution for the period of this study.

Cases of suspected child abuse were prospectively identified before the postmortem examination. The diagnosis of SBS was made by the multidisciplinary Suspected Child Abuse and Neglect team using at least two of the following criteria: (1) typical abnormal findings on neuroimaging, (2) typical skeletal injury, (3) retinal hemorrhages, (4) history of abusive shaking with or without blunt head trauma, or (5) an inadequate history to explain the observed injuries. In all cases of suspected SBS, one or more of the authors (C.S., R.W.E., or A.V.L.) performed modified orbital exenteration at the postmortem examination. All histologic slides were reviewed by at least two of the authors (C.S., A.V.L., or A.S.).

At The Hospital for Sick Children, it has been the standard procedure of at least one pathologist (C.S.), in children who die from accidental trauma, to examine orbits for optic nerve sheath hemorrhage by blunt dissection from above after the orbital roof is removed. Exenteration was performed only if optic nerve sheath hemorrhage was suspected macroscopically by the presence of a bluish tinge to the optic nerve sheath. Only the cases performed by this pathologist were included in this study. The number of cases performed by other pathologists during the duration of this study is unknown and unavailable.

This new autopsy technique was designated to be best practice (and continues as our routine approach) for all cases of suspected inflicted childhood neurotrauma (SBS with or without impact) starting in 1989. We retrospectively identified all cases between 1989 and 2000, a period that coincidentally yielded a similar number of cases of accidental head trauma that had also received exenteration by the investigating coauthor pathologist (C.S.).

This study was conducted before our institution required Research Ethics Board approval for retrospective reviews. However, it was conducted in a fashion consistent with current guidelines.

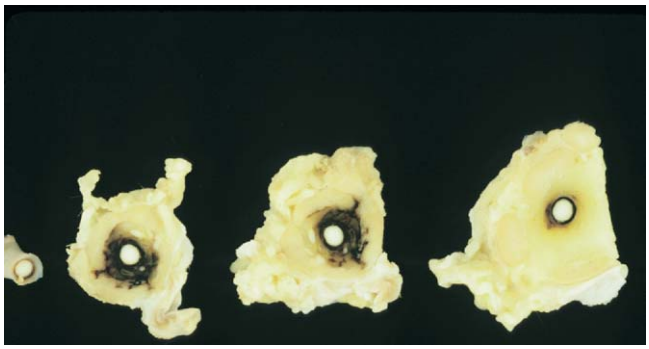

**FIGURE 1. Serial section of the orbit in Shaken Baby Syndrome (SBS). Anterior orbit to the right, canalicular optic nerve to the left. Note hemorrhage within optic nerve sheath and extending into the orbital tissues surrounding optic nerve in the posterior orbit.**

• **TECHNIQUE FOR ORBITAL EXENTERATION AT POSTMORTEM:** The vitreous was not routinely aspirated for toxicology studies by the pathologist before exenteration.

As the brain was removed, the optic nerves were severed just posterior to their exit from the optic canal into the cranial vault. The orbital roof was then removed piecemeal without damaging the underlying periosteum using a striker/electric saw, large scissors, bone forceps, or rongeur. The optic canal was then unroofed using the rongeur. In SBS cases, the periosteum was bluntly dissected away from the orbital walls and severed at the margins of the inferior orbital fissure in a manner that also severed nerves and vessels entering the fissure. In accidental head trauma patients, the superior aspect of the periosteum was incised to allow gentle, blunt exposure of the optic nerve. If optic nerve sheath hemorrhage was suspected, then the remainder of the periosteum was dissected from the orbit as described.

After performing a 360-degree conjunctival peritomy and undermining Tenon fascia, the anterior part of the orbit was liberated by cutting the capsulo-palpebral ligaments and lower lid retractors, inferior oblique at its proximal origin, and superior oblique tendon in the area of the trochlea. After careful retraction of the eyelids, often with the use of a lid speculum, the orbital septum was severed from its attachment to the orbital rim, thus establishing a connection to the intracranial portion of the orbit. The entire globe and orbital contents, enclosed by periosteum, along with the canalicular optic nerve were then removed via the intracranial space *en bloc*. The specimen was fixed in buffered formaldehyde for 48 to 72 hours and then sectioned for examination.

The globe was then separated from the optic nerve by coronal section using a hand-held blade. Axial sections of the globe were examined including the macula and the

**TABLE 1.** Ocular Signs of Injury in Shaken Baby Syndrome and Accidental Trauma

| Injury | Shaken Baby Syndrome | Accidental Trauma | 95% CI* of Risk Difference |
|---|---|---|---|
| Retinal hemorrhages | 16/18 (89%) | 3/18 (17%) | (0.50–0.95) |
| Any optic nerve hemorrhage | 14/18 (78%) | 7/18 (39%) | (0.09–0.69) |
| Optic nerve subdural hemorrhage | 14/18 (78%) | 6/18 (33%) | (0.15–0.73) |
| Optic nerve intradural hemorrhage | 11/18 (61%) | 2/18 (11%) | (0.23–0.77) |
| Optic nerve epidural hemorrhage | 5/18 (28%) | 3/18 (17%) | (–0.16–038) |
| Optic nerve subarachnoid hemorrhage | 5/18 (28%) | 0 | (0.07–0.48) |
| Hemorrhage within orbit: around optic nerve anterior | 12/18 (67%) | 4/18 (22%) | (0.15–0.73) |
| Hemorrhage within orbit: around optic nerve posterior | 7/18 (39%) | 1/18 (6%) | (0.08–0.58) |
| Hemorrhage within orbit: around optic nerve anterior and posterior | 6/18 (33%) | 1/18 (6%) | (0.04–0.52) |
| Hemorrhage within orbit: around optic nerve anterior, mid, and posterior | 5/18 (28%) | 1/18 (6%) | (–0.01–0.45) |
| Hemorrhage within orbit: around optic nerve mid and posterior | 1/18 (6%) | 0 | (–0.05–0.16) |
| Orbital fat hemorrhage | 9/18 (50%) | 4/18 (22%) | (–0.02–0.58) |
| Extraocular muscle hemorrhage | 5/18 (28%) | 0 | (0.07–0.48) |

*CI = confidence interval.

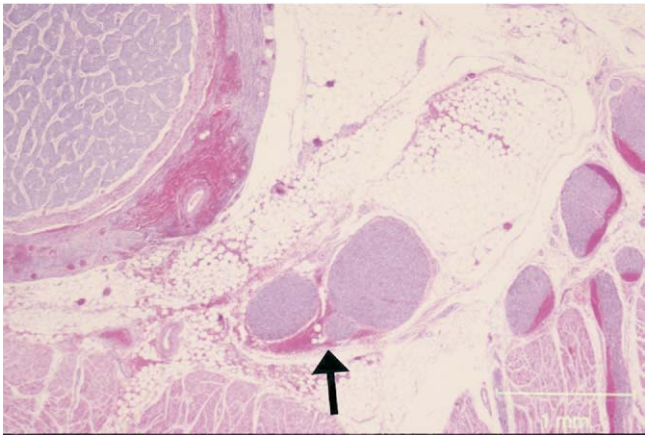

**FIGURE 2. Histologic section near orbital apex from child victim of Shaken Baby Syndrome (SBS). Note hemorrhage within dura, orbital fat, and in the sheath of the cranial nerves (arrow). [Hematoxylin and eosin]**

optic nerve. Coronal sections of the orbital contents were taken sequentially in an anteroposterior direction (Figure 1) and examined by light microscopy. Signs of ocular and intracranial trauma were noted; in particular, hemorrhage.

For the purpose of this study, we reviewed all available microscopic sections of the eyes and orbits from study subjects. Signs of injury to the scalp, skull, and intracranial contents were recorded from the autopsy report. History of the trauma mechanism in accidental cases was extracted by review of the medical records. Stains for hemosiderin were not routinely performed and therefore the data are not included here. Likewise, we did not routinely record data regarding signs of inflammatory response or lymphocytic infiltration, although this was not a predominant finding in any case.

After cutting the pupil-optic section, the pathologist conducted gross examination of both calottes noting the extent and location of retinal hemorrhages. These data are not presented in the present study.

• **STATISTICAL ANALYSIS:** We examined associations between the SBS group and the accidental trauma group with regard to epidural optic nerve hemorrhage and location of the hemorrhage in the anterior, mid, or posterior orbit in addition to orbital fat hemorrhage. The second association was between subdural optic nerve hemorrhage, intradural optic nerve hemorrhage, epidural, or subarachnoid optic nerve hemorrhage. The third association performed was between intradural optic nerve hemorrhage and subarachnoid optic nerve hemorrhage and, finally, orbital fat hemorrhage was correlated to extraocular muscle hemorrhage or epidural hemorrhage.

• **DATA ANALYSIS:** SAS Institute Software for windows version 9.1 (SAS Institute Inc, Cary, North Carolina, USA), was used for statistical analysis of the data. *P* values less than 0.05 were regarded as statistically significant. We used the Fisher exact test with permutation *P*-value adjustment for multiple test procedure.

## RESULTS

BETWEEN 1989 AND 2000, POSTMORTEM ORBITAL EXENTERation was performed in 36 children. Eighteen were victims of SBS and 18 died as a result of accidental head trauma. Mean patient age in the SBS group was 13.3 months old (range, one month to three years) and in the accidental trauma group five years (range, two days to 14 years). Male to female ratio in the SBS group was 2:1 and in the accidental trauma group was 1:2.8.

Optic nerve sheath hemorrhage was found in 14/18 (78%) children in the SBS group, The optic nerve hem-

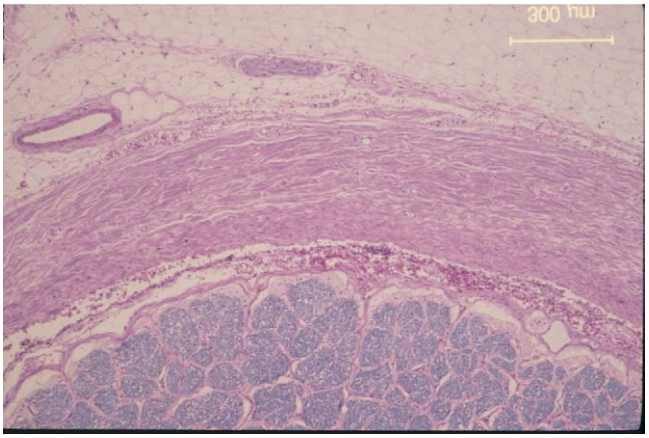


**FIGURE 3. Histologic section near orbital apex from child victim of motor vehicle accident. Child ejected approximately 50 feet from vehicle and landed head first. Note scarcity of hemorrhage within dura and immediately epidural. [Hematoxylin and eosin]**

**TABLE 2.** Signs of Head Trauma in Shaken Baby Syndrome and Accidental Trauma

| | Shaken Baby Syndrome (n = 18) | Accidental Head Trauma (n = 18) |
|---|---|---|
| Any sign of head trauma | 15 (83%) | 18 (100%) |
| Subdural CNS hemorrhage | 17 (94%) | 12 (67%) |
| Subarachnoid hemorrhage | 12 (67%) | 13 (72%) |
| Cerebral edema | 14 (78%) | 14 (78%) |
| Tonsillar herniation | 10 (59%) | 8 (42%) |

CNS = central nervous system.

orrhage was subdural in all 14 children, intradural in 11 (61%), subarachnoid in five (28%), and epidural (immediately surrounding and external to the nerve sheath) in five (28%). The optic nerve sheath hemorrhage was located in the anterior orbit in 12 of 18 (67%) cases, mid orbit in five (33%), and posterior orbit in seven (39%). In some cases, it was in two or more of these locations. Five (28%) children demonstrated a continuous hemorrhage within the entire optic nerve sheath (Table 1).

In the accidental trauma group, optic nerve sheath hemorrhage was observed in seven of 18 (39%) cases. The hemorrhage was subdural in six (33%, $P = .0176$), intradural in two (11%, $P = .0045$) with 95% confidence interval for risk difference (0.07–0.48), and epidural in three (17%, $P = .6906$). Subarachnoid hemorrhage was not observed in any of the children in this group ($P = .0455$). Only two (11%) patients had optic nerve sheath hemorrhage occupying more than one-third of the orbital length (Table 1).

Only one SBS case did not exhibit hemorrhage beyond the epidural area of the optic nerve. Epidural hemorrhage was located only in the anterior orbit in five of 18 (28%) cases. In the accidental trauma group, epidural hemorrhage was located only in the anterior orbit in four (22%, $P = .0176$) cases; this was significantly lower than in the SBS group in the anterior, mid, and posterior orbit in one (6%, $P = .1774$) case and in the posterior orbit only in one case (6%, $P = .0408$). Only one patient had epidural optic nerve hemorrhage throughout the orbit. SBS children had a significantly higher rate of subdural hemorrhage ($P = .0176$).

Hemorrhage into the extraocular muscles was seen in five of 18 (28%) of the SBS patients. This finding was not identified in any of the accidental trauma patients ($P = .0455$). One SBS victim had hemorrhage in the cranial nerve sheaths (Figure 2). This was not specifically looked for in all patients; therefore, the frequency of this finding in both groups is unknown.

Epidural orbital fat hemorrhage, and orbital hemorrhage extending well beyond the epidural area with involvement of extraocular muscle was observed in nine of 18 (50%) of the SBS cases (Figures 1 and 2). It was more severe in the posterior orbit in most cases (Figure 1). In the accidental head trauma patients, hemorrhage into orbital fat was seen in only four of 18 (22%) cases and the blood was epidural or in the region of orbital fracture. This difference was not statistically significant compared with the SBS group ($P = .1642$). However, when the three cases of orbital fracture were removed from the accidental trauma (no orbital fractures were seen in the SBS group) group, hemorrhage into the orbital fat was only seen in two of 15 (13%) of the patients. The difference was of statistical significance ($P = .0488$ with a confidence interval for risk difference being 0.08–0.65).

The combination of orbital and optic nerve sheath hemorrhages were highly significantly more common in the SBS group ($P < .001$). Diffuse optic nerve hemorrhages in all layers including epidural optic nerve hemorrhages occurring together were not more common in the SBS group even when omitting the three cases with fractures of the orbit. However, multilayered optic nerve sheath hemorrhages excluding epidural involvement were statistically more common in the SBS children ($P < .001$).

Retinal hemorrhages were observed in 16 of 18 (89%) cases of SBS. In the accidental trauma group, retinal hemorrhages were observed in only three of 18 (17%, $P < .0001$). In two of these three cases, the hemorrhages were superficial intraretinal, and in the remaining case, the hemorrhages were dot intraretinal. Despite the fatal nature of the trauma, in all three cases, the hemorrhages were few in number and confined to the posterior pole. All three accidental trauma cases with retinal hemorrhages involved severe acceleration-deceleration injury. The second and third cases described in the following section were also two of the three cases who had orbital fat hemorrhage in the accidental trauma group. The first case was a two-year-old child who was sitting in a restraint seat of a car that rolled

over in a motor vehicle accident. He was thrown at least 20 m from the vehicle and found face down. He sustained multiple cranial and cerebral injuries. There was no evidence of injury to organs of the chest or abdomen. He had a scanty optic nerve sheath hemorrhage with microscopic evidence of a very small number of red blood cells in the dura and the epidural tissues (Figure 3). In the second case, a seven-year-old boy was crushed by a falling boulder and suffered from lacerations of the brain parenchyma, cerebral contusion and cerebral edema, and direct injury to the orbit, which included orbital fat hemorrhage. In the remaining case, a 12-year-old girl was riding a horse and fell. Her foot got entangled in the stirrup while the horse continued to gallop, dragging her head hitting the ground repeatedly. When the horse finally stopped the child was dead. The latter two cases had more orbital hemorrhage than did the first case.

Intracranial signs of injury were observed in all victims of SBS (Table 2). Subdural hemorrhage was observed in all cases, subarachnoid hemorrhage in 11 of 17 (65%), cerebral edema in 12 of 17 (71%), and tonsillar herniation in 10 of 17 (59%). Signs of blunt head trauma were observed in 15 of 17 (88%) cases: nine (53%) with contusion of the brain and six (35%) with fractures of the skull. In the accidental trauma group, cerebral subdural hemorrhage was present in 12 of 19 cases (63%), subarachnoid hemorrhage in 13 of 19 (68%), edema in 13 of 19 (68%), and tonsillar herniation in eight of 19 (42%). Signs of head trauma were identified in all patients in keeping with the inclusion criteria for this group (Table 2). No patient had subgaleal hemorrhage. We also performed the association between the entire accidental trauma group, and thereafter excluded those three cases in which the accident resulted in orbital fractures (Table 3).

## DISCUSSION

WE REVIEWED THE ORBITAL PATHOLOGY OF A CONSECUtive cohort of child victims of SBS and compared them with a cohort of children who died as a result of accidental head trauma.

The reported postmortem ophthalmic findings in SBS have largely been confined to ocular and optic nerve pathology. Lambert and associates first described the presence of optic nerve hemorrhage in 1986.[10] Subdural optic nerve sheath hemorrhage has since been observed in 65% to 100% of shaken babies.[11–14] The most common site for optic nerve sheath hemorrhage is in the immediate retrobulbar portion of the optic nerve.[6] Other reported orbital findings include the presence of intradural optic nerve sheath hemorrhage,[11,12] and intrascleral hemorrhage noted at the junction of the sclera and the optic nerve.[13,15]

The technique of *en bloc* orbital exenteration was developed by us following the finding of a posterior optic nerve sheath laceration in the orbit of the first SBS child on whom we elected at autopsy to explore the orbital tissues in 1989 (Figure 4). At that time, the orbital anatomy was not preserved, but we recognized that orbital trauma was potentially occurring in this patient group. As a result, the technique was modified to allow intact *en bloc* orbital exenteration. This procedure also allows viewing of the victim in the funeral home with an undisturbed facial appearance.

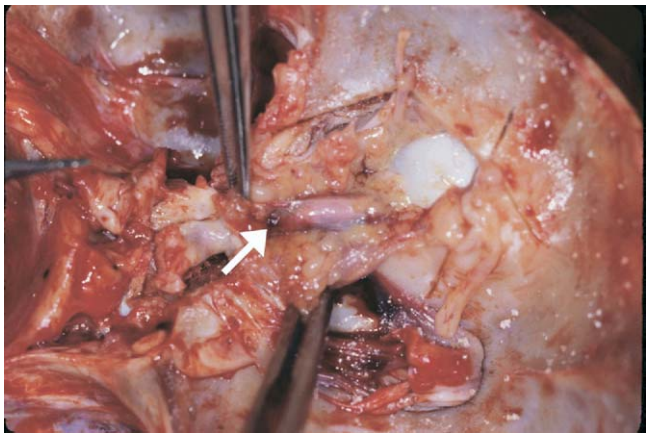

**FIGURE 4. View of unroofed orbit in a child victim of Shaken Baby Syndrome (SBS). Posterior aspect of the globe is seen from above *in situ* facing to the right. Note bluish discoloration of optic nerve sheath from subdural blood. Forceps is holding tissue just posterior to a laceration (arrow) in the dural sheath.**

We found that optic nerve sheath hemorrhage in fatal accidental blunt head injury was significantly less common than in SBS (39% vs 78%; $P < .04$). Because we only conducted orbital exenteration on cases in which gross inspection of the optic nerve sheath led to a suspicion of hemorrhage within the sheath, and because we found at histology a far lower incidence than this gross inspection had predicted, then the overall incidence of optic nerve sheath hemorrhage in accidental head trauma is likely to be even lower. We believe it is unlikely that cases with no gross evidence of hemorrhage would have blood seen on histology. Nevertheless, we note that there was a procedural difference between the two groups.

Only three children with accidental head trauma had orbital hemorrhage, one of whom had sustained direct blunt trauma to the orbit (boulder fell on head); in these three cases, the physical damage to the children was dramatically more severe. The incidence of orbital hemorrhage in SBS was much higher (94% vs 16%). Orbital fat hemorrhage was seen in 50% of SBS victims and only 22% of accidental head injury (the three cases in which there was orbital hemorrhage). In one of the accidentally injured children (motor vehicle accident), the amount of hemorrhage in the orbit and dura was miniscule, being visible only histologically and even then of minimal quantity. Extraocular muscle hemorrhage occurred only in SBS victims.

**TABLE 3.** Statistical Significance of Results Excluding Orbital Fracture Cases in Accidental Trauma Group

| Variable | Raw *P* Value | Adjusted *P* Value |
|---|---|---|
| Retinal hemorrhages | 0.00002 | .00 |
| Any optic nerve hemorrhage | 0.01267 | .030 |
| Subdural optic nerve hemorrhage | 0.01267 | .030 |
| Intradural optic nerve hemorrhage | 0.00140 | .00 |
| Epidural optic nerve hemorrhage | 0.28322 | .720 |
| Subarachnoid optic nerve hemorrhage | 0.03610 | .170 |
| Orbital fat hemorrhage | 0.02993 | .150 |
| Subarachnoid edema | 0.13281 | .515 |
| Optic nerve epidural hemorrhage in any location, or orbital fat hemorrhage | 0.000071 | .000 |
| Extraocular muscle hemorrhage | 0.03610 | .170 |
| Any optic nerve hemorrhage | 0.012670 | .010 |
| Any optic nerve hemorrhage except epidural hemorrhage | 0.000286 | .000 |
| Orbital fat or extraocular muscle or epidural hemorrhage | 0.002541 | .005 |

*P*-values adjusted for multiple testing.

In the SBS group, the optic nerve hemorrhage involved several levels of the optic nerve sheath, and occupied a larger portion along the length of the orbit. Our study is consistent with previous series demonstrating that subdural optic nerve sheath hemorrhages are common in SBS.[11,12] In our series, they were observed in all cases with optic nerve hemorrhage. In the SBS group, 79% had intradural optic nerve sheath hemorrhage, which was observed in only 18% of the accidental trauma group; this difference was statistically significant ($P < .0045$). In keeping with other reports, we found that intradural hemorrhages in SBS are most common at the optic-scleral junction.[11] SBS victims (28%) had subarachnoid optic nerve hemorrhage that was not seen in any of the accidental trauma victims, yet this is less common than subdural and intradural optic nerve hemorrhages. Only 28% of the SBS victims demonstrated orbital hemorrhages confined to the anterior orbit. The majority of children had more extensive orbital hemorrhage, with the most severe hemorrhage always seen in the posterior orbit. Only one child in the accidental trauma group had diffuse orbital hemorrhage, which occurred after direct orbital injury from a boulder.

Our findings suggest that orbital and optic nerve injury is more common and more severe in SBS than accidental head trauma, especially when located in the posterior orbit. We only observed orbital injury in accidental head trauma when there was direct trauma to the orbit accompanied by orbital fractures or severe multiple repetitive acceleration deceleration (motor vehicle rollover, girl dragged by horse). This leads us to suspect that there is a unique feature of repeated acceleration-deceleration injury, as seen in SBS and our accidental cases, which leads to orbital trauma. The increased frequency of hemorrhage at the anterior and posterior portions of the optic nerve sheath and orbit suggest that the junction of the globe and optic nerve and the firm posterior orbital attachments of all orbital structures sets up fulcrums across which force is generated during acceleration deceleration, leading to tissue damage at these locations. In addition, SBS children attain a significantly higher rate of diffuse orbital hemorrhage, including the optic nerve, orbital hemorrhage without optic nerve involvement, and, finally, deep optic nerve hemorrhages are also more prevalent in this group. This supports the preliminary computer and animal modeling results of Parsons and associates (Parsons MA, Cirovic S, Bhola RM, et al: A finite element model of the eye and orbit to stimulate nonaccidental injury in infants. North American Conference on Shaken Baby Syndrome, Montreal, Canada, September 13 to 14, 2004). They suggest that the orbital fat moves with the globe as if there was a suction coupling attachment.

The cause of subdural or subarachnoid optic nerve sheath hemorrhage in SBS is unknown. Lambert and associates suggest that an acute rise in intracranial pressure, which is transmitted to the optic nerve sheath causing expansion of the sheath with rupturing of bridging veins may play a role.[10] Rapid rises in intracranial pressure in adult and animal models can be associated with optic nerve sheath and retinal hemorrhage.[16] Although this may indeed explain optic nerve sheath hemorrhage in SBS, it does not account for the presence of intrascleral,[13] intradural, or orbital hemorrhage. Increased intracranial pressure does not correlate with retinal hemorrhages in SBS.[17] It has also been suggested that optic nerve sheath, intradural, retinal, and perhaps even orbital hemorrhage, are due to obstruction to venous outflow from the retina and orbit. The rarity of a clinical appearance of central or branch retinal venous obstruction and the lack of correlation with increased intracranial pressure (the mechanism by which the retinal venous pressure would hypothetically rise) in SBS makes this hypothesis difficult to support. In addition, the facial and pterygoid venous systems provide alternative drainage, are not valved, and are not affected by increased intracranial pressure.[18] Although the blue tinge that led us to suspect optic nerve sheath hemorrhage in the accidental trauma cohort was not well correlated with the true presence of bleeding, it may have represented venous engorgement. Yet, it was apparently insufficient to commonly result in hemorrhage despite the severity of the injuries.

Another theory proposes that optic nerve sheath subdural hemorrhage (SDH) or subarachnoid hemorrhage (SAH) represent direct extension of intracranial hemorrhage. In Terson syndrome, intracranial, particularly subarachnoid hemorrhage, is associated with retinal and vitreous hemorrhage.[18,19] However, the hypothetical mechanism of direct extension of blood is not supported by postmortem findings in SBS reported here and by others,

in which it is found that the optic nerve sheath hemorrhages are predominantly retrobulbar, often discontinuous, and without blood along the proximal part of the optic nerve sheath as the optic nerve passes through the optic canal.[6,20] In addition, it has been shown that there is no correlation between the sidedness of intracranial hemorrhage and the sidedness of retinal hemorrhage.[17] Schloff and associates found that Terson syndrome in children is uncommon. They predicted a maximal incidence of intraretinal hemorrhage in patients with nonabuse intracranial hemorrhage to be 8%.[21]

Other authors have suggested that intrascleral hemorrhage in SBS may be caused by acceleration-deceleration trauma that results in tears of the intrascleral ciliary vessels forming the circle of Zinn-Haller.[13,15,22,23] Previously reported patients with intrascleral hemorrhage at the junction of the optic nerve and sclera have not demonstrated signs of direct blunt trauma to the globe. Immunohistochemical staining studies of postmortem specimens with markers such as β-amyloid precursor protein also identify this site as a prime area for nerve damage in SBS.[24,25] Thus a combination of mechanical trauma and vascular insult, perhaps in both the anterior and posterior orbit, may lead to the optic atrophy, which is the second most common cause of visual loss in SBS survivors.[7] The most common cause is occipital cortical damage. Indeed, there has been no other viable explanation for optic atrophy in SBS to our knowledge.

The orbital optic nerve is S-shaped and has some inherent redundancy because it is longer than the distance from the orbital apex to the eyeball. The intraorbital portion of the optic nerve is 25 mm to 30 mm long. The optic foramen and the insertion of the nerve into the eye are, on average, only 18 mm apart. The eye is also easily translated in the orbit. These factors may allow the globe to move in the orbit at the time a child is shaken, or accidentally submitted to severe repeated acceleration-deceleration forces, thus applying forces to the optic nerve and orbital structures, which are fixated at the orbital apex and the globe, creating loci for injury at these two points.

We remain puzzled about the relationship between orbital injury and retinal hemorrhages. There is extensive evidence that links severe hemorrhagic retinopathy in otherwise well children to SBS as opposed to accidental head injury.[7] This implicates acceleration-deceleration forces and vitreo-retinal interface shearing as important factors. The unique presence of traumatic retinoschisis in SBS supports this conclusion. Perhaps orbital trauma directly disrupts vascular supply to or from the retina. We are particularly intrigued by the finding of cranial nerve sheath hemorrhage in one patient and regret that we did not evaluate this finding further in other patients. The cranial nerves carry autonomic nervous supply to the globe. The autonomic nervous system is involved with vascular autoregulation. Autoregulation of retinal vessels can be disrupted by shearing forces in the cat model.[26] Dysregulated vessels become more patulous and prone to leakage. It has been theorized that the retinal hemorrhages seen in normal newborns may be due to prostaglandin effects, a chemical also involved in autoregulation of retinal vessels.[27]

The presence of orbital pathology seems to be a consistent finding in the SBS cases we studied using this new technique for postmortem ocular and orbital examination. We are aware that some of the children examined in the accidental trauma group were older than the SBS group, which may make them biologically different. This is a difficult problem to resolve because life-threatening accidental head trauma is rare in the nonaccidental trauma age group. Further use of this technique is recommended to assist in the better understanding of the ocular injuries induced by nonaccidental repeated acceleration deceleration, the etiology of retinal hemorrhages, and the forensic significance of our findings.

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## Biosketch

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## Biosketch

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