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Ophthalmology of shaken baby syndrome

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Ophthalmology consultation is a critical part of the evaluation of a child who may be a victim of shaken baby syndrome (SBS) as retinal hemorrhages are a cardinal manifestation of this form of child abuse. Although it was Caffey who first recognized that retinal hemorrhage was a manifestation of violent shaking [1,2], many previous reports exist that in retrospect clearly demonstrate this association [3,4]. Over the last 35 years there has been an expansion of research and knowledge regarding the pathophysiology and specificity of retinal hemorrhage as they relate to SBS. Different patterns of retinal hemorrhaging may have different levels of specificity and different mechanisms of causation. But increasingly it is recognized that shaking itself is the key element in generating hemorrhagic retinopathy.

Anatomy

Not all retinal hemorrhages are the same. It is absolutely essential that the term is not used generically; no more so than one would use the word "fracture" to describe all fractures. Retinal hemorrhages should be described in terms of number, patterns of distribution, and type. Such distinctions may have important forensic and diagnostic implications.

The retina is a multilayered structure that lines the inside of the eyeball. Its edge, called the ora serrata, is found just behind the iris. As a result, the peripheral retina lies out of the view of the direct ophthalmoscope thus making ophthalmology consultation, with use of pharamacologic pupillary dilation and indirect ophthalmoscopy, essential in order to view the entire retina. The direct ophthalmoscope is only able to view the posterior pole; defined as the optic nerve, the retina immediately surrounding the optic nerve (peripapillary retina), the macula, and the fovea (Fig. 1). The superior and inferior temporal vascular arcades that emanate from the optic nerve delimit the macula. The fovea is the area at the centre of the macula specialized to provide optimum acuity for straight-ahead viewing.

The location of a retinal hemorrhage within the retina will determine its appearance. Hemorrhage in the superficial nerve fibre layer will take on a somewhat linear appearance: a splinter or flame shaped hemorrhage. Retinal hemorrhages located deeper in the retinal tissue will take on a round or amorphous shape called a dot or blot hemorrhage. Although blot hemorrhage is a term used to refer to a hemorrhage bigger than a dot, there are no strict measurements for this distinction. Hemorrhage in front of the retina, lying on its surface (preretinal hemorrhage), is characterized by the obscuration of underlying retinal vessels that course through the superficial retinal layers. Likewise, vessels running over the hemorrhage characterize subretinal hemorrhage. Blood can also spread into the vitreous gel that fills the eye behind the iris (vitreous hemorrhage) or it can become trapped between the vitreous and retina (subhyaloid hemorrhage).

The vitreous of young children is much more adherent to the retina than it is in adults. These attachments are greatest at the macula, over retinal blood vessels, and at the peripheral retina. Shaking of the child will set the vitreous into motion causing shearing forces to be applied to the retina via these attachments. As a result, the retina of the macula

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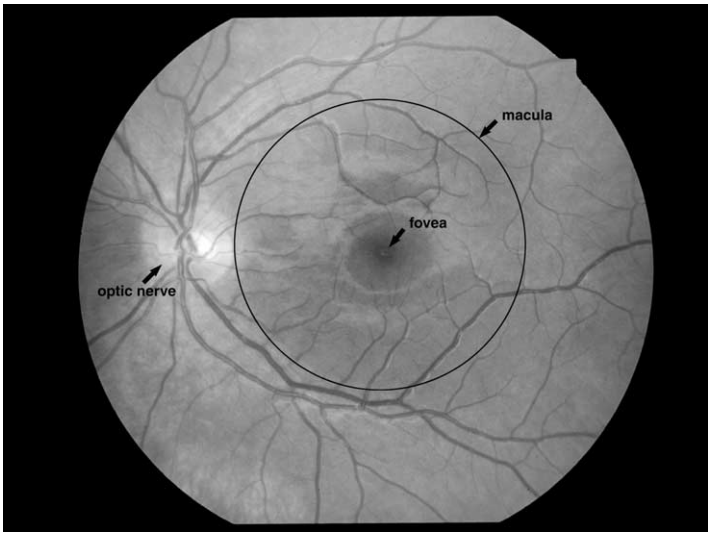

Fig. 1. Normal retina and optic nerve. Everything seen in this image would be considered part of the posterior pole.

may be split apart at any of its 10 layers with blood accumulating in the intervening cystic cavity. This lesion is called traumatic retinoschisis [5,6] and may have been described in shaken babies before the term was coined [7]. Although adults may present with lesions that on first glance look similar [8], in children in the SBS age range, it has never been observed in any circumstance other than SBS. In these adults, the cause is readily apparent and the anatomy different [8]. When present in children less than 4 or 5 years old, it is a very specific indicator that shaking has occurred. Although the visual prognosis is usually good, blood can break out of the cavity into the vitreous. It may be difficult to distinguish blood within a traumatic retinoschisis from preretinal or subhyaloid hemorrhage. The distinction can clearly be made by the recognition of circumlinear edges of the affected retina that may be hemorrhagic or hypopigmental and raised as the retina is folded by the pull of the vitreous [9]. Either acutely or later after healing, hypopigmentation may be seen at this edge (Fig. 2). Ophthalmology consultation can be essential in recognizing these important findings due to the ability of the ophthalmologist to get a more global view of the affected area as opposed to the limited view of the direct ophthalmoscope.

Perhaps also due to vitreous traction, peripheral retinal hemorrhage can be another indicator that shaking is likely to have occurred [10–12] . Although this is also a location often affected in leukemia and hypercoagulopathies, it is an area that is very infrequently involved when retinal hemorrhage is associated with most other etiologies such as accidental head trauma (see below).

Other patterns of retinal hemorrhage may occur in shaken babies but may also be suggestive of particular causes other than shaking. Hemorrhage confined to a perivascular distribution may indicate vasculitis or coagulopathy. Hemorrhage radiating out from the optic nerve and involving the entire retina in this centripetal pattern is characteristic of central retinal vein occlusion, an uncommon manifestation of SBS which may be seen in a variety of uncommon pediatric medical conditions.

Papilledema is a common cause of retinal hemorrhage. These hemorrhages are located on the optic nerve head or in the immediately adjacent retina and mostly appear as splinter hemorrhages radiating out from the swollen disc. It is important to note that papilledema does not cause hemorrhages elsewhere in the retina. In fact, in children in the SBS age range (≤3 years old with rare exception [10]), elevated intracranial pressure is also only a very rare cause of hemorrhagic retinopathy. Hemorrhages due to papilledema are entirely non-specific and reflect whatever caused the optic nerve swelling whether it is intrinsic optic nerve disease, orbital compression, or intracranial pathology. Hemorrhage from optic nerve swelling is not indicative of SBS although it may be seen in this disorder. Papilledema is actually uncommon in SBS [13].

Approximately two thirds of shaken babies will show severe hemorrhagic retinopathy: too

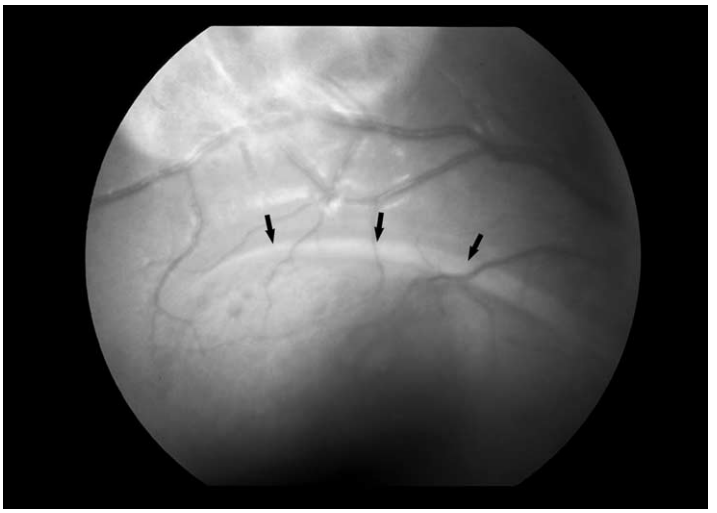

Fig. 2. Traumatic retinoschisis. Note circumlinear hypopigmented retinal fold (*arrows*) at edge of cavity over which blood vessels bend.

numerous to count hemorrhages; pre-, intra-, and subretinal hemorrhage; and distribution throughout the retina to the ora serrata [13,14]. However, some victims of SBS may have fewer hemorrhages confined only to intraretinal distribution in the posterior pole. In fact, on average approximately 15%–25% have no hemorrhages at all [10,13,15]. Hemorrhage may also be asymmetric between the two eyes in terms of number, type or distribution. Unilateral hemorrhagic retinopathy may also occur [10,13,16].

Pathophysiology

Recognition of traumatic retinoschisis as a unique feature of SBS informs us that repetitive acceleration-deceleration forces play a key role in generating retinal injury. This type of traction-shearing may also explain the prevalence of peripheral retinal hemorrhage in SBS as the vitreous is also well attached in this area. These findings have led researchers to call previous theories into question [10]. In the past, it has been suggested that the presence of intracranial hemorrhage (Terson syndrome), increased intracranial pressure, or increased thoracic pressure (as might be induced when the perpetrator grabs a child by the chest) play a role in generating retinal hemorrhage. Although there may be cases in which each of these findings may play some role, it appears that shaking itself is the important factor.

Terson syndrome, the combination of intracranial bleeding with retinal and vitreous hemorrhage is not uncommon in adults. Work in our center has shown that in shaken babies there is no correlation between intracranial hemorrhage of any particular type and retinal findings. Likewise, our prospective evaluation of children with causes of intracranial hemorrhage other than abuse found retinal hemorrhage to be rare; and when it occurs, the hemorrhages are few in number and confined largely to the posterior pole. It is important to note for those that propose theories relating to facile rebleeding of subdural hematomas, that even if this were to occur, the blood itself (from the first bleed or the rebleed) is an insufficient explanation to account for anything more than a few hemorrhages in the posterior pole. However, it has been suggested that perhaps the presence of a chronic effusion, supported by the presence of pre-injury macrocephaly, could rarely make a child more susceptible to acceleration-deceleration as the force is propagated more easily through the fluid [17,18]. One author reports a single case of a child with macrocephaly who fell from standing against a piece of furniture and sustained a subdural hemorrhage and retinal hemorrhage. However, the author notes that the bilateral subhyaloid, intraretinal, and preretinal hemorrhages are out of proportion to that ever reported, only hand drawn pictures are provided, and several other details are missing

from the account. The hemorrhages appear confined to the posterior pole. Although dilation of the subarachnoid spaces around the optic nerves is suggested as playing a role, this is not documented.

The pathophysiologic mechanisms of Terson are unknown. Blood in the optic nerve sheath is thought to be an important factor. Some have suggested that blood can track directly into the optic nerve sheath from the brain. One study attempts to show a correlation between sidedness of retinal hemorrhages and intracranial blood but the study number was so low (3 cases) that the observation could easily have been due to chance [19]. When we conducted a similar statistical analysis on a larger group of patients (76), there was no correlation. In addition, shaken babies with retinal hemorrhage may or may not have optic nerve sheath hemorrhage and even when present; it may not involve the entire course of the nerve [10,20].

The evidence that suggests that it is shaking itself that is the primary cause of retinal hemorrhage continues to mount [5,9,10,12,14,15,21,22]. In addition to the vitreal traction and direct retinal shearing or contusion from the vitreous [15], it appears that as the whole globe is shaking, orbital structures are also moving. The optic nerve is tethered at its entry to the sclera and at the apex. Hemorrhage may be observed at these sites both within the optic nerve sheath, the sclera [20], the apical orbital fat, and the sheaths of nerves running through the orbit [10]. I am aware of one case of optic nerve avulsion due to shaking (personal communication, Dr. Suzanne Starling) and one case in which postmortem dissection at our centre revealed a laceration of the posterior optic nerve sheath. Staining for β-amyloid precursor protein supports the presence of diffuse axonal injury, as seen in the brains of shaken babies, also in the optic nerves of shaken babies but not in controls or a single case of accidental head trauma that was tested. Our research shows that SBS orbital injuries are only seen in accidental head trauma when the acceleration-deceleration forces are extremely severe and/or repetitive. It may be that injury to the autonomic supply to the eye has a role in disruption of vascular integrity and autoregulation leading to retinal hemorrhage in SBS. Direct injury to the optic nerve may explain the presence of optic nerve sheath hemorrhage (rather than Terson syndrome) and the not uncommon optic atrophy, which is the second most common cause of visual loss in survivors.

Timing

Retinal hemorrhages can not be dated. The variability in the timing of their resolution is dramatic and unpredictable. Although it is reasonable to state that flame and small intraretinal hemorrhages usually resolve faster than preretinal and larger hemorrhages, and that the blood in a traumatic retinoschisis cavity can take weeks or months to clear, the ranges of timing are so large that this information is rarely useful in identifying the time of the injury. Certainly there may be outlier situations that defy rationale explanation. One would never expect a flame hemorrhage to last for months. Findings that are indicative of an ''earlier event'', as measured in weeks or months rather than a few days, would include optic atrophy, retinal scarring, or fixed folds in a detached retina. It is sometimes tempting for the ophthalmologist to suggest the occurrence of two separate shaking events based on differences of colour and density of hemorrhages. This too should be avoided. It must also be stressed that the timelines offered below for resolution of birth hemorrhages do *not* apply to SBS, as the mechanism of injury is different. For example, local shearing damage to the retina in SBS may influence the ability of a hemorrhage to resolve as compared to what appears to be an otherwise healthy retina following birth.

Differential diagnosis

Severe hemorrhagic retinopathy in the SBS age range is almost always due to SBS (Fig. 3). Those disorders that may mimic SBS are usually easy to diagnose either due to the pattern of hemorrhages or the presence of obvious systemic or laboratory abnormalities. For example, leukemia can be diagnosed by routine blood testing. Leukemic retinopathy is often characterized by the presence of white leukemic infiltrates and an increased number of peripheral hemorrhages due to sludging of blood flow in the presence of very high leukemic cell counts. Even the most severe coagulopathies rarely cause severe hemorrhagic retinopathy. Perhaps the only exception is neonatal vitreous hemorrhage due to congenital protein C deficiency; an entity that has a clearly defined systemic presentation that would not likely be confused with SBS [23]. The more mild elevations in measures of clotting status, some of which may be seen in association with head injury [24], would not be expected to cause much in the way of retinal hemorrhaging.

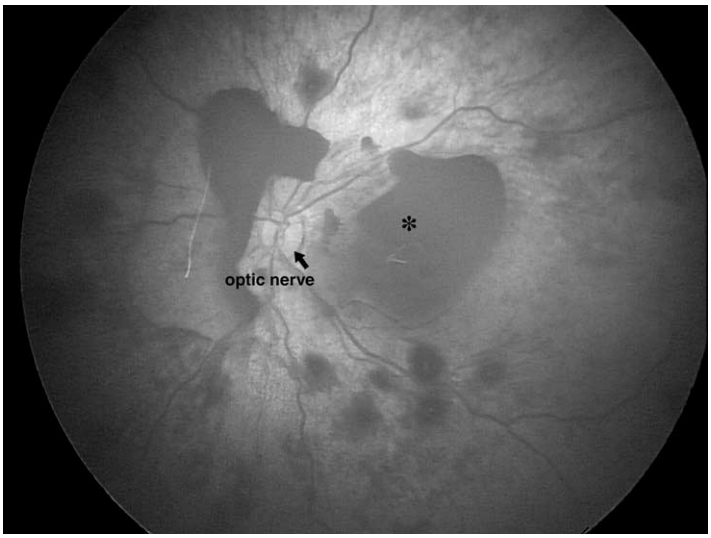

Fig. 3. Severe hemorrhagic retinopathy due to shaken baby syndrome demonstrating large areas of preretinal and subhyaloid hemorrhage (*) as well too-numerous-to-count flame and dot hemorrhages. No papilledema.

Yet, it is advisable that coagulation status be measured in all children in which hemorrhage is felt to be a possible manifestation of abuse. Vasculitis can also cause retinal hemorrhages but the perivascular pattern is usually diagnostic. Exudate may also be present.

A wide variety of primary retinal diseases may also cause hemorrhaging but these disorders should be readily recognized by an experienced ophthalmologist. For example, retinopathy of prematurity has a very characteristic appearance with hemorrhages arising from the neovascular vessels on the peripheral ridge which demarcates the edge of vascularized retina. Of course these children also have a history of being born prematurely. There is nothing about their retina that would otherwise make them predisposed to hemorrhages elsewhere or at a later date when the disease is no longer active. Congenital infectious retinopathies such as those caused by cytomegalovirus or herpes simplex, have typical appearances including the presence of exudate and necrotic retina often confined to specific geographic areas. Once again, diagnostic systemic, laboratory, and/or radiologic signs are likely to be present.

Lesser hemorrhagic retinopathy, a small number of intraretinal (and perhaps preretinal) hemorrhages confined to the posterior pole, is a much less specific diagnostic indicator (Fig. 4). Yet, even if one considers the long list of possible causes [10], two things become clear: (1) even when hemorrhage does occur it is usually very uncommon, and much less common than retinal hemorrhage due to SBS, and (2) the reported cases are characterized by very few hemorrhages in the posterior pole. This interpretation can be applied to retinal hemorrhage due to anemia, elevated intracranial pressure (ICP), glutaric aciduria, cardiopulmonary resuscitation and many other diagnoses. It may be the presence of retinal hemorrhages characteristic of SBS that allows one to ascertain that a systemic disorder, which has never previously been reported as a cause of such hemorrhages, is not the cause of the child's acute decompensation because indeed, the child has been shaken [25].

Of particular interest to neurosurgeons is the potential for elevated intracranial pressure to cause retinal hemorrhaging. In adults, acute rises in ICP have been shown to be associated with blood in the optic nerve sheath [26]. There are some authors who suggest that elevated ICP may play a role in causing the retinal hemorrhages of SBS but their theories no longer seem applicable in light of more recent findings [5,27,28]. For example, Munger felt that the presence of choroidal and scleral hemorrhage could not be explained by vitreous traction and therefore suggested elevated ICP as the cause. Today, the importance of orbital shaking is being recognized: a mechanism that would explain both of these findings. In a review of 76 cases of SBS, we were

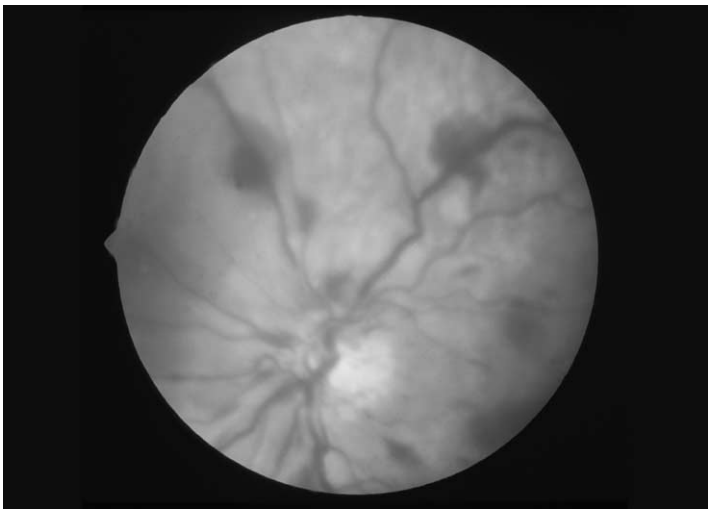

Fig. 4. Mild hemorrhagic retinopathy due to shaken baby syndrome.

unable to find any statistical correlation between elevated ICP and retinal hemorrhaging. However, rare cases of central retinal vein occlusion in SBS may be due to elevated ICP, but this pattern of hemorrhaging in the retina is very specific with hemorrhages radiating out in a spoke-like fashion throughout the retina from the optic nerve. The central retinal vein could also be injured directly by orbit shaking. Otherwise, in children, retinal hemorrhages from elevated ICP are very uncommon and when they do occur, usually present as no more than a few pre- or intraretinal hemorrhages around or on the optic nerve head. The authors of one large postmortem study concluded that brain "edema alone is not sufficient to cause retinal hemorrhages or other (intrascleral, optic nerve sheath) ocular hemorrhages" [12]. Although not formally studied prospectively, thousands of nonabused children have been examined by ophthalmologists in the presence of even severe acute increased intracranial pressure. To my knowledge, severe hemorrhagic retinopathy has never been reported.

There are also uncommon reports suggesting that retinal hemorrhages in children could be secondary to aneurysm, arteriovenous malformation, or arachnoid cysts. In all of these entities, in children, retinal hemorrhages are only very rarely present and when they are, they tend to be few in number, intra- or preretinal, and confined to the peripapillary retina or the optic nerve itself [10, 29,30]. One must be careful to distinguish hemorrhages due to papilledema, which may be common in these entities from unrelated retinal hemorrhages [10,31]. There is one case of "extensive bilateral retinal hemorrhages and a large right subhyaloid hemorrhage" in a 6 week old child with an aneurysm confirmed by angiography [32]. It is noted that there were significant psychosocial and historical risk factors for abuse, the neuroimaging was not consistent with SBS, and no investigation for abuse was conducted.

Some disorders that commonly cause retinal hemorrhage in adults never cause this finding to occur in the SBS age range. These include diabetes and sickle cell or other hemoglobinopathies. Although hypertension is a frequent cause of retinal hemorrhage in adults, it is an extremely rare cause in children and even then almost always recognizable by the presence of retinal exudation, systemic abnormalities, and of course, high blood pressure. Bacterial endocarditis can uncommonly cause white centered hemorrhages (Roth spots) in adults. I am unaware of any reports of this occurring in the SBS age range. In fact, white centered hemorrhages are entirely non-specific as they can be seen with virtually any cause of retinal hemorrhage. They may be due to septic emboli, central fibrin, central resolution, cellular infiltrate, or even light reflection from the examining source.

Perhaps the only cause of retinal hemorrhages in young children that exceeds SBS in frequency would be normal childbirth. Up to 50% of babies will be found to have hemorrhages in the first 36 hours of life [10,33]. This drops off rapidly to 11%–15% by 72 hours after birth [10].

Hemorrhages can be found following any type of delivery although they are more common after spontaneous and vacuum assisted vaginal delivery [33]. Thousands of babies have been studied from around the world giving us solid data to rely on regarding these figures. Although the hemorrhagic retinopathy of normal birth may be quite dramatic with too-numerous-to-count hemorrhages, and involvement out to the ora, preretinal hemorrhage is uncommon, subretinal hemorrhage is rare and retinoschisis never observed [10,33,34]. Vitreous hemorrhage is also rare [33]. These babies also have none of the other sequelae of shaking such as intracranial hemorrhage and fractures. The retinal hemorrhages characteristically resolve without any long term effect; thus the reason why babies are not routinely examined by ophthalmologists. Flame hemorrhages are always gone by the end of the first week of life and dot/blot hemorrhages by 6 weeks (usually by 4 weeks) with rare exception only in extraordinary cases [10,35]. Hemorrhages of these types within these time guidelines may be indistinguishable from SBS. However, should hemorrhages of these types occur outside of these guidelines, then they should not be attributed to birth [15].

Severe life threatening accidental head trauma, on the scale of a motor vehicle accident or a fall from a great height, may cause a mild hemorrhagic retinopathy in less than 3% of cases [10,16,36–41] . In fact, most studies show a zero incidence. It may be that unique severe accidents of great magnitude may be able to generate enough acceleration-deceleration force to yield a minimal representation of the severe repetitive acceleration-deceleration that characterizes shaking. Minor household trauma such as falls from sofas, out of the arms of caretakers, and during play rarely if ever cause retinal hemorrhages [10,41,42]. It took one author a study of tens of thousands of records to find just 4 children who had retinal hemorrhages as a result of playground falls, two of whom fell off of moving swings [43]. Even in a study that carries a title suggesting that household falls might rarely cause retinal hemorrhage, the mechanism of injury was far beyond that which might be labeled mild or common (eg, a fall through a banister from the top of a flight of stairs head first onto a concrete floor) [40]. Even when the uncommon event of retina hemorrhage due to accidental head trauma does occur, the hemorrhages are usually few in number, mostly confined to the posterior 50% of the retina, and intra- or preretinal. As with SBS and birth, unilaterality or asymmetry may occur. Retinoschisis has never been reported following accidental injury in the SBS age range and subretinal hemorrhage is rare. However, if there is also direct blunt trauma to the eye during the injury, more serious damage may be seen.

The lessons learned from the many articles dealing with the usual absence of retinal hemorrhage following accidental head trauma suggest that the retina follows the common laws of tissue injury as observed in other causes of trauma such as burns (Fig. 5). Presumably, there is a threshold for tissue damage that would result in retinal hemorrhage. Extremely severe forces, above those that a human being can generate, can allow this threshold to be reached even when delivered over a very short time period. However, in order for human forces to be able to reach the tissue damage threshold, the force (in this case acceleration-deceleration) must be delivered over a longer period of time. This is supported by the confessions of perpetrators who describe repeated and sustained shaking during the incident of injury.

As rib fractures can be seen in shaken babies, the question has been raised regarding the possibility of inducing retinal hemorrhage via the chest compressions of cardiopulmonary resuscitation (CPR). Extreme increases in intrathoracic pressure can be transmitted to the retina via the venous system. The resultant retinal appearance, originally recognized by Purtscher a century ago, is characterized by white retinal patches, the pathophysiology of which remains obscure, along with a paucity of hemorrhages. These patches can occasionally be seen in SBS [44]. In one small series, all children with Purtscher retinopathy following severe chest crush injuries had unilateral retinal hemorrhage, optic atrophy, and white patches [45]. But Purstcher retinopathy has never

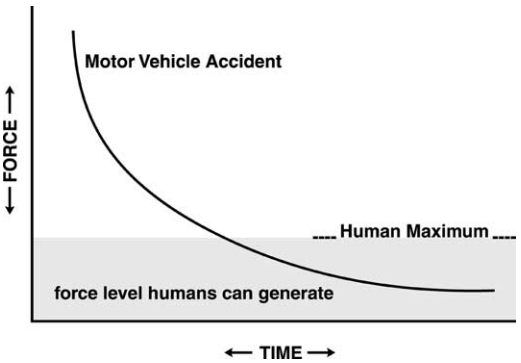

Fig. 5. Tissue damage curve. Injury (retinal hemorrhages) occurs on the right side of the curve.

been described following CPR despite multiple studies in children [10,46]. In fact, retinal hemorrhage following CPR appears to be extremely rare if at all possible. Despite several studies [10,47–49] there is only one convincing case in the literature in which a baby without obvious other predisposing factors such as coagulopathy, who had been examined before and after CPR, had a single retinal hemorrhage in one eye [50]. Clearly, CPR is not a satisfactory explanation for moderate or severe hemorrhagic retinopathy. If one considers that even severe traumatic chest compression (e.g. motor vehicle accident) has never been reported in young children to cause a hemorrhagic retinopathy, it seems unreasonable to think that improperly applied CPR by a lay person could result in retinal hemorrhages.

Occasionally, it will be suggested that a caretaker shook a child in an attempt to resuscitate them. Although the forces which cause SBS are not completely understood, we know from the confessions of perpetrators that it is an extremely violent act that lies beyond that which a well meaning caretaker might use while attempting to arouse a child. Yet, perhaps in an extremely unusual circumstance, such as a single reported case of an extreme household fall combined with shaking to resuscitate, hemorrhages could occur [42]. In this case, the bilateral hemorrhages were not further detailed. The story of well-intended resuscitation may actually be a cover for the true abusive event [7]. There is a correlation between the severity of the brain injury in SBS and extent of the retinal hemorrhages [13,51].

Other eye injuries due to shaking

Although uncommon, severe shaking can lead to disruption of the intraocular contents with retinal detachment or avulsion usually with giant tears [52], hyphema, cataract, and glaucoma. Inflammatory sequelae have also been reported in these severely damaged eyes [52]. More commonly, although still seen in only a minority of cases, is retinal edema. This may occur as a result of local impairment of blood flow or it may indicate direct blunt trauma to the eye (retinal commotio, Berlin edema). Strangulation or suffocation may result in subconjunctival hemorrhage. Ocular signs secondary to the brain injury may include pupillary abnormalities and cranial nerve palsies. Peripheral retinal scarring has been described in the French literature [52,53]. The lesions tend to be located in the far temporal periphery with a round shape and irregular edges. It is unclear whether these result from retinal hemorrhage or perhaps, shearing forces elicited by the peripheral retinal attachments to the vitreous.

Prognosis

Retinal hemorrhages rarely result in damage to the retina that is visually significant. Even macular traumatic retinoschisis can settle with good visual outcome [5]. However, if vitreous hemorrhage or foveal retinal hemorrhage occurs, then deprivation amblyopia may result particularly when the eyes are not affected equally. For this reason it is important that children with traumatic retinoschisis or large areas of preretinal hemorrhage be followed carefully for these complications. There is one reported case in which the traction induced retinoschisis was so severe that vascular avulsion and retinal ischemia was induced leading to neovascularization of the optic nerve head [6].

Despite the usual resolution of retinal hemorrhage without sequelae, 15%–28% of SBS survivors will be blind [54,55]. The most common causes of visual loss are central nervous system cortical damage (by infarction, contusion, atrophy, cystic encephalomalacia, or laceration) and optic atrophy. The latter is not due to retinal hemorrhage and probably reflects the direct injury to the optic nerve in the orbit during shaking as discussed above. Strabismus may also be seen as a result of neurologic or visual compromise [52].

Management

Whenever there is any suspicion of possible SBS or when a child in the SBS age range dies suddenly and without explanation, ophthalmology consultation is indicated. This should include pharmacologic dilation of the pupils except in those circumstances where the child's moribund status already renders the pupils fixed and dilated. Unfortunately, there is often a disagreement between neurosurgeons and ophthalmologists about dilating drops. If the neurosurgeon feels strongly that constant preservation of pupillary reactivity is absolutely essential to the child's neurologic management then other options may be considered. Short acting mydriatics such as phenylephrine 2.5% can return pupillary activity to normal within 4–6 hours. If one eye is dilated at a time, this allows for continuous observation for an afferent pupillary defect (Marcus Gunn pupil) [56]. Ophthalmologists may also have viewing

systems that allow for visualization of the retina through small pupils. But it must be stressed that pupil saving techniques such as those described here may yield a suboptimal examination that could limit the ability of the ophthalmologist to form an educated opinion regarding the retinal findings. Wherever possible, bilateral simultaneous full pharmacologic pupillary dilation is preferred.

It is essential that documentation be complete. Our studies and observations by others [7,13] have shown that although non-ophthalmologists are often correct in their identification of the presence or absence of retinal hemorrhages, false positives and false negatives do occur. In addition, our research shows that non-ophthalmologists rarely if ever state in their notes any description of the hemorrhages preferring instead to simply state that they are there.

It is incumbent upon consulting ophthalmologists to describe their findings in details, draw accurate pictures, and if possible, photograph the abnormal retina. There are many imaging systems available, all of which are limited by the need for some technical training and high cost [57]. Images may also be limited by contrast, vitreous hemorrhage, angle of view, small pupil size, and patient compliance [14]. Yet when available and possible, photographs offer a component of documentation that can be very useful. Photography should never replace the live examination by an ophthalmologist as photographs may miss important diagnostic signs, often afford limited view of the peripheral retina, and render what the ophthalmologist sees in binocular 3 dimensions as a flat 2-dimensional image. There may also be legal issues with regards to chain of custody and the potential for altering digital images. It is recommended that the flow of images from the camera at the bedside to the database or chart be strictly documented and tracked.

There is no treatment for retinal hemorrhages. Although there may be some thought given to evacuating blood from a traumatic retinoschisis cavity, this will usually resolve spontaneously and due to layering, rarely causes a prolonged obstruction of the visual axis. I am aware of cases in which surgery has been attempted, yet found to be very difficult as the macular retina may be further injured and the blood difficult to extract. The use of laser has been suggested to liberate a subhyaloid collection of blood in an adult [8], but I am not aware of this being applied to the SBS population. Surgery may be useful in cases of non-resolving vitreous hemorrhage that are blocking vision and therefore likely to result in amblyopia. Patching of the better eye is the mainstay of amblyopia treatment and should be considered by the attending ophthalmologist whenever there is unequal obstruction to the visual axis between the eyes. Long term follow-up is also essential to look for visual sequelae that may not be apparent in the intensive care unit such as cortical visual impairment and optic atrophy.

Outliers

Like most medical conditions, there continue to be cases and reports in the literature which seem to represent situations that lie outside of the current boundaries of scientific understanding. For example, an article in the neurosurgical literature suggests that simple falls of young Japanese children on tatami mats can result in subdural and retinal hemorrhage [58]. Why does this article report an event that clearly runs contrary to virtually everything we know about retinal hemorrhages? Although the reader is referred elsewhere for a more detailed analysis [10], it is important to note that the article was published at a time when the particular country of origin had no clear laws or investigation teams to consider child abuse. Some articles in the literature may report events that occurred before the recognition of SBS as an entity in the early 1970s [4]. In fact, even Caffey who eventually was credited with being one of the first to recognize SBS as a form of child abuse, had an earlier publication in which the mechanism of retinal and intracranial hemorrhage was not fully appreciated [3]. Other papers suffer from a lack of sufficient detail, failure to recognize and/or exclude other possible causes of retinal hemorrhage where such etiologies should have been considered, or most commonly a failure to provide an adequate description of the retinal hemorrhages. It is this latter problem that most disturbingly leads to conclusions about ''retinal hemorrhages'' as if all were the same. This illustrates the critical importance of obtaining ophthalmology consultation and describing the amount, distribution, and type of hemorrhages in all cases. It must also be remembered that those children who have other conditions that might explain their retinal hemorrhages are not immune from being shaken as well.

One area that remains puzzling is the possibility of retinal hemorrhages without intracranial hemorrhage. Three articles report such a situation, although in two cerebral edema was present, in one an MRI demonstrated hemorrhage that was not

apparent on CT, and all were from the earlier years of neuroimaging [59–61]. Other authors have mentioned this possibility in passing without case evidence [49]. Cases of severe intracranial hemorrhage with little or no retinal hemorrhaging as well as the converse are well recognized. Cerebral edema alone, when seen in other situations, does not appear to be a cause of severe hemorrhagic retinopathy. Perhaps some forms of shaking induced shearing from acceleration-deceleration can rarely affect the eyes without causing bleeding in or around the brain.

Ongoing research at our center continues to explore the pathophysiology and differential diagnosis of retinal hemorrhages. Animal models may also be useful. For example, our work on woodpeckers has showed that they may be protected from ocular injury by the presence of anatomic features which restrict the ability of the globe to shake within the orbit as well as intrascleral bone and cartilage which renders the sclera less deformable. Yet despite the continuous need for further investigations, we must also listen to the ever-increasing body of knowledge that tells us that retinal hemorrhage, particularly when severe, likely indicates the child has been shaken.

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