Optical Coherence Tomography Findings in Shaken Baby Syndrome

VEIT STURM, KLARA LANDAU, AND MARCEL N. MENKE

• PURPOSE: **To evaluate features of optical coherence tomography (OCT) associated with Shaken Baby syndrome (SBS) in an attempt to gain new insight into the pathophysiology of this phenomenon.**
• DESIGN: **Institutional prospective observational case series.**
• METHODS: **We report three infants with SBS. Each patient underwent an ophthalmic examination including slit-lamp biomicroscopy, dilated indirect ophthalmoscopy, color fundus photography, and OCT.**
• RESULTS: **In all infants, numerous bilateral multilayered retinal hemorrhages were present. In one case, bilateral perimacular folds had occurred. OCT confirmed retinal hemorrhages and perimacular folds. Moreover, OCT revealed vitreoretinal traction in all infants and suspected hemorrhagic macular retinoschisis in one case. Based on OCT findings, a hypothesis of vitreoretinal traction development and retinal fold formation is proposed.**
• CONCLUSIONS: **OCT provided valuable additional information about the ocular pathology in patients with SBS. Vitreoretinal membrane formation seen in OCT could support the pathophysiological theory of a direct mechanical effect. OCT revealed preretinal blood accumulation as a cause for localized vitreous detachment and vitreoretinal traction. Furthermore, OCT showed persistent attachment of the vitreous to the internal limiting membrane at the apices of perimacular folds and suggested small hemorrhagic macular retinoschisis in one patient. Perimacular folds and hemorrhagic macular retinoschisis are regarded as highly specific for SBS and indicate poor visual outcome. Thus OCT might be of both diagnostic and prognostic value in SBS. (Am J Ophthalmol 2008;146:363–368. © 2008 by Elsevier Inc. All rights reserved.)**

TRAUMA IS THE MOST COMMON CAUSE OF DEATH in childhood, and nonaccidental head trauma in infants—the so—called Shaken Baby syndrome (SBS)—is the leading cause of infant death from injury.[1,2] Crying is a major factor in inciting SBS.[3,4] The characteristic findings that suggest inflicted head trauma in SBS consist of subdural, and/or subarachnoidal hemorrhages, extensive retinal hemorrhages. and encephalopathy in infants.[5] Pediatric ophthalmologists are regularly asked to examine the retina of children suspected of having been abused, because retinal hemorrhages have been found in 83% of affected children.[6]

Two major theories on the cause of retinal hemorrhages exist: The first postulates raised retinal venous pressure attributable to sudden increases in chest or head pressure. The second theory of vitreoretinal traction from repeated acceleration-deceleration postulates a direct mechanical effect of the shaking or impact itself.

Optical coherence tomography (OCT) was introduced in 1991 as a noncontact, noninvasive biomedical imaging technique.[7] OCT uses a scanning interferometer that produces high-resolution (approximately 10 μm longitudinal resolution) cross-sectional images of retinal layers. In 1995, OCT was used first for imaging macular diseases.[8,9] This technology has been useful in detecting and following various retinal disorders, such as macular holes, macular edema, intraretinal thickening, epiretinal traction, pigment epithelial detachment, choroidal neovascular membranes, subretinal fluid, and macular traction.[10–13] OCT can also identify nerve fiber loss in glaucoma and other optic nerve disorders.[14,15]

We studied OCT features associated with SBS in an attempt to gain new insight into the pathophysiology of this phenomenon.

METHODS

THREE INFANTS WITH SUSPECTED SBS WERE REFERRED TO our institution within one year (November 1, 2006 to October 31, 2007) for ocular examination. Their ages ranged from four to seven months. Patients were prospectively included in this observational case series.

Each patient underwent an ophthalmic examination including slit-lamp examination, dilated indirect ophthalmoscopy, color fundus photography, and OCT. For that purpose, the OCT machine was moved to the Pediatric Intensive Care Unit. OCT scans were obtained while the patients were sedated because of their general condition. Infants were placed in an anti-Trendelenburg position in front of the OCT machine being held by a second person. The maximum height of the OCT chin rest was increased

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From the Department of Ophthalmology, University Hospital of Zurich, Zurich, Switzerland.
Inquiries to Veit Sturm, Department of Ophthalmology, University Hospital of Zurich, Frauenklinikstrasse 24, CH-8091 Zurich, Switzerland; e-mail: veit.sturm@usz.ch

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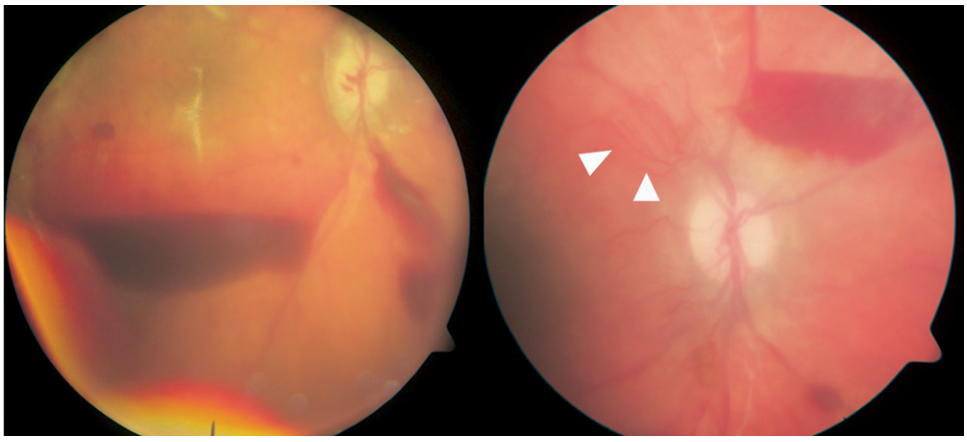

FIGURE 1. Fundus images of the right eye of Patient 1 with Shaken Baby syndrome (SBS). (Left) Severe hemorrhagic retinopathy showing large preretinal and subhyaloidal hemorrhages as well as flame and dot hemorrhages. (Right) Three weeks later, subtotal resorption of hemorrhages and newly developed intraretinal neovascularization (arrowheads) is seen.

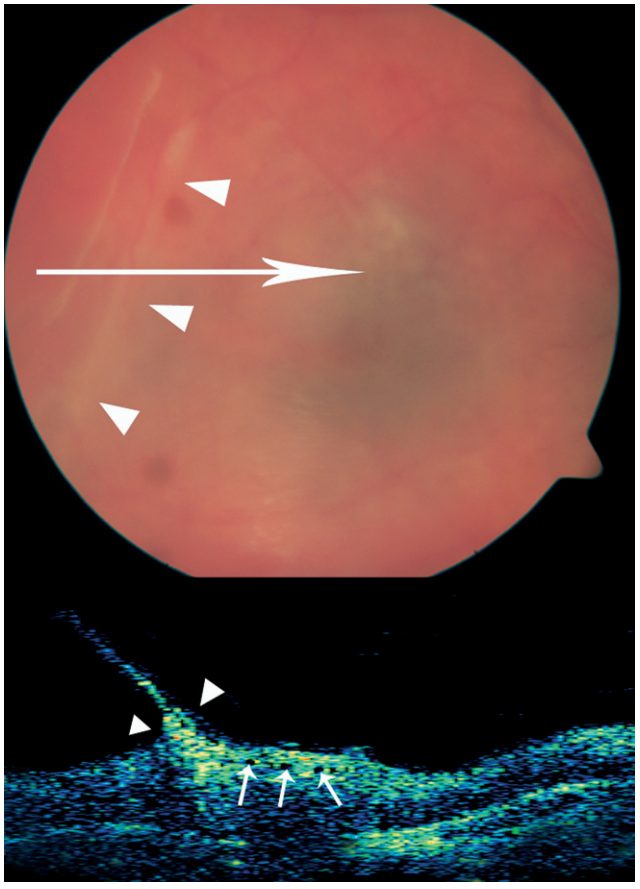

FIGURE 2. Fundus image and corresponding optical coherence tomography (OCT) findings of Patient 1 with SBS. (Top) Hypopigmented perimacular retinal folds can be seen (arrowheads). The arrow indicates scan location and direction of the OCT scan. (Bottom) Corresponding OCT scan showing the retinal fold (arrowheads) with persistent attachment of the vitreous. Arrows indicate schisis cavities.

by a gel pad usually used for positioning support in surgical settings. The eyelids were kept open by a lid speculum. OCT images were recorded after dilated examination of the fundi. Multiple cross-sectional images of the retina were obtained using the fast macular thickness protocol of a Zeiss Stratus OCT3 (Carl Zeiss Meditec, Dublin, California, USA). The fast macular scan was selected to maximize imaging ability in patients naturally unable to maintain fixation on a target. Whenever possible, higher resolution line-scans were performed as well. OCT images were only recorded from the more affected eye as derived from findings using indirect ophthalmoscopy. As patients were unable to fixate on the fixation target, and a foveal reflex could not be seen in the OCT infrared fundus image, the anatomic relationship of the optic disk and vascular arcades was used as a landmark for OCT imaging.

RESULTS

• **PATIENT 1:** A seven-month-old healthy boy with epileptic seizures was transferred to the Pediatric Intensive Care Unit. The patient was intubated and conventionally ventilated for seven days. Clinical and computed tomography (CT) examination revealed a skull fracture, subdural hemorrhage with diffuse brain edema, and a hematoma of the neck. Three days after initial hospitalization, the child suffered from a stroke.

Ophthalmological examination revealed bilateral multilayered flame-shaped retinal hemorrhages and perimacular folds. The diffuse pre-/intraretinal and subhyaloidal hemorrhages were located predominantly in the posterior pole but extended also to the ora serrata. The right eye was more affected (Figure 1, Left). Within three weeks, the large preretinal hemorrhage along the inferior arcade resolved but extensive peripapillary neovascularizations developed. (Figure 1, Right). Traumatic retinoschisis with

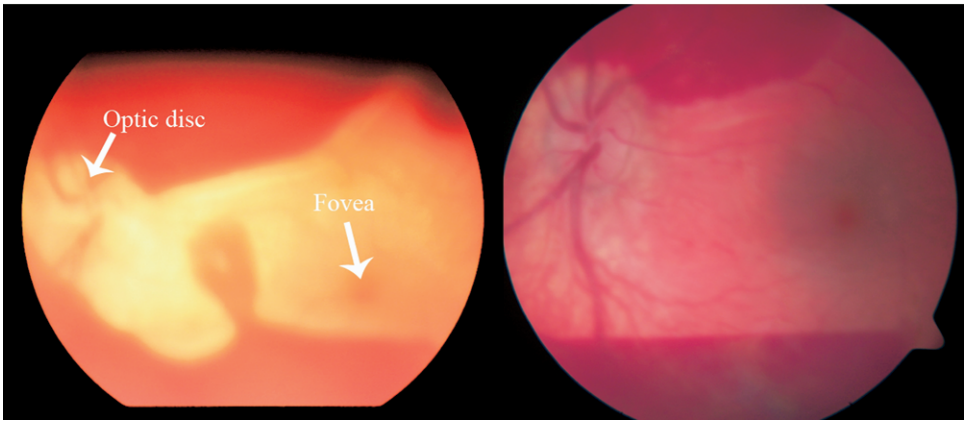

FIGURE 3. Fundus images of the left eye of Patient 2 with SBS. (Left) The fundus image shows extensive retinal and preretinal hemorrhages around the optic nerve head covering the lower and upper retina and relatively sparing the macular region. (Right) Four weeks later, hemorrhages had resorbed partially.

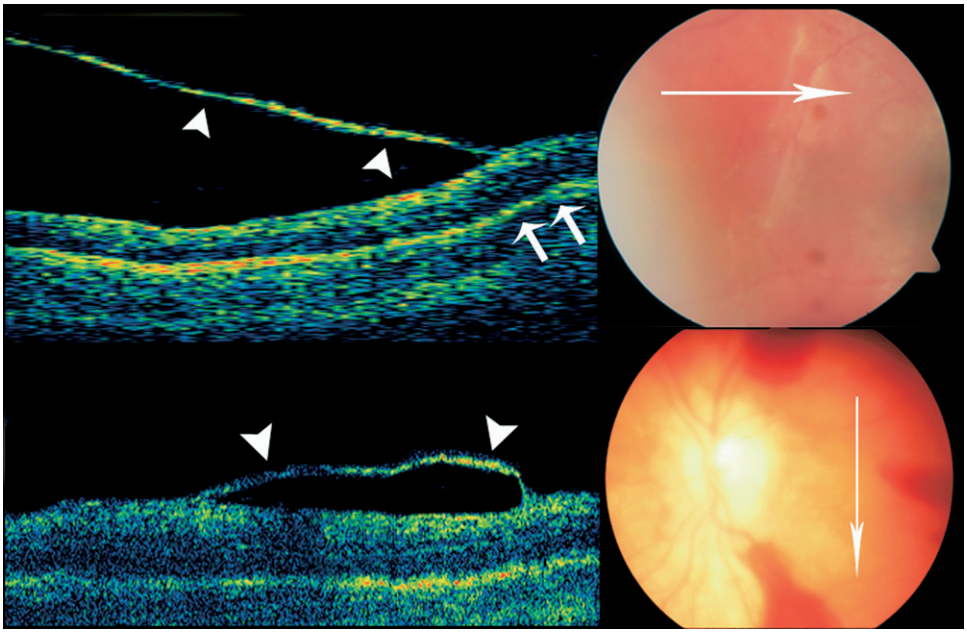

FIGURE 4. Vitreoretinal traction and epiretinal membrane (ERM) formation in SBS. (Top left) Horizontal OCT scan in Patient 1. Extensive vitreoretinal traction can be appreciated (arrowheads) pulling up the retina causing retinal fold formation (arrows). (Top right) Corresponding fundus photograph indicating the scan location at the edge of the fold. (Bottom left) OCT scan of Patient 3 showing ERM formation. (Bottom Right) Corresponding fundus image demonstrating the scan location.

hypopigmented retinal folds at the edge of cavity persisted (Figure 2). The father's confession was obtained.

• **PATIENT 2:** A four-month-old boy was reported by the partner of the mother to have suffered a seizure with consecutive brief loss of consciousness. Upon admission to the Pediatric Intensive Care Unit, a bulging fontanel and hypothermia were noted. CT and magnetic resonance imaging (MRI) showed acute subdural hematoma in the left temporo-occipital region. The patient suffered recurrent seizures.

Fundus examination of the right eye showed extensive retinal and preretinal hemorrhages around the optic disk covering the lower and upper retina, but relatively sparing the macular region. The left fundus was more affected (Figure 3, Left) with a subretinal hemorrhage in the lower retina of the left eye. Four weeks later, the bleeding was only partially resorbed (Figure 3, Right). The confession of the mother's partner was obtained.

• **PATIENT 3:** A five-month-old boy was brought to the the Pediatric Intensive Care Unit because of suspected

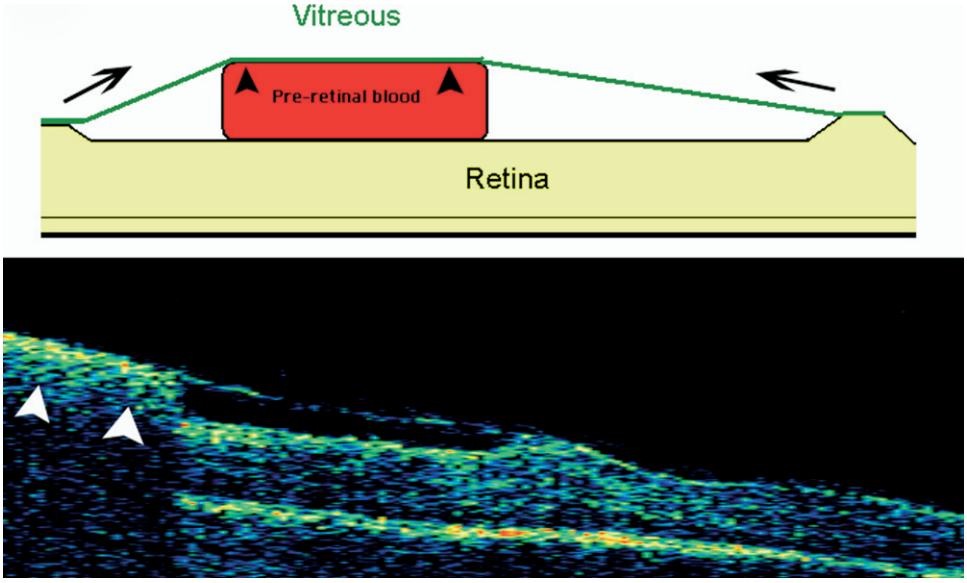

FIGURE 5. Mechanism of vitreoretinal traction and retinal fold formation in SBS. (Top) Scheme demonstrating the possible mechanism of vitreoretinal traction and retinal fold formation attributable to pre-retinal blood accumulation. The mechanism might be similar in cases of hemorrhagic retinal pigment epithelium detachments. Preretinal blood applies pressure on the vitreous body leading to indentation and localized vitreous detachment with consecutive vitreoretinal traction in the surrounding area. (Bottom) Corresponding OCT findings in Patient 2 supporting this hypothesis.

foreign body aspiration. Cardiopulmonary resuscitation had to be performed. The child was intubated and conventionally ventilated for three days. On closer examination, the boy had bilateral orbital hematomas. CT showed bilateral frontal subdural hygroma with right-sided hemorrhage. A cerebrospinal fluid drainage was performed. The boy suffered recurrent seizures.

Furthermore, ophthalmological examination showed posterior pole flame-shaped retinal hemorrhages as well as numerous isolated and confluent dot hemorrhages in the mid-peripheral retina extending to the ora serrata similar as in Patient 2 (Figure 4, Bottom right). The left eye was even more affected.

The diagnosis of SBS was established based on the combination of fundoscopic and CT findings.

• **OPTICAL COHERENCE TOMOGRAPHY FINDINGS:** Multiple serial OCT scans from the central retina could be obtained. OCT confirmed multilayered macular hemorrhages in all patients. The perimacular folds seen by indirect funduscopy in Patient 1 were affirmed by OCT (Figure 2, Bottom). In addition, OCT showed perimacular retinal traction not noted in fundoscopy in all patients (Figures 4 and 5) and suspected small hemorrhagic macular retinoschisis in Patient 1 (Figure 2, Bottom). The morphology of the neurosensory retina in close distance to retinal folds was severely disturbed. Therefore, exact localization of schisis cavities was not possible. However, cystic cavities were mostly seen in the inner retinal layers and might have even extended into the nerve fiber layer.

DISCUSSION

TO OUR KNOWLEDGE, OCT FINDINGS HAVE NOT YET REported in SBS. OCT is not routinely being used in children. In a previous report, OCT was performed under general anesthesia in a child with nystagmus.[16] An alternative approach for obtaining an OCT using a simple sedation with propofol has been suggested.[17] We examined infants in an anti-Trendelenburg position in front of the OCT while they were sedated because of their general condition. Multiple serial OCT fast-macular-scans were performed to evaluate the contour of the central region of the retina.

Optical coherence tomography scans were unable to detect the foveal pit. This might be attributable to the severe morphological changes associated with SBS, such as retinal hemorrhages and retinal edema. In addition, patients were unable to cooperate and examination time had to be reduced to a minimum in these critically ill infants. OCT confirmed retinal hemorrhages in all patients.

Previously a direct mechanical effect has been postulated leading to retinal changes associated with SBS. Massicotte and associates reported persistent attachment of the vitreous to the internal limiting membrane at the apices of perimacular folds and suggested that this finding might constitute evidence of violent shaking.[18] OCT affirmed perimacular folds. In fact, with OCT we could demonstrate such a persistent attachment of the vitreous at the apices of perimacular folds (Figure 2, Bottom). OCT showed vitreoretinal traction already seen on fundoscopy

(Patient 1). In addition, OCT demonstrated extensive vitreoretinal traction in Patients 2 and 3, which was not seen in ophthalmoscopy attributable to obscuration by severe hemorrhages. Those images demonstrating vitreoretinal traction could be reproduced using scans in various directions. The near infrared light of the OCT seems to be less disturbed by vitreous hemorrhage compared with conventional indirect ophthalmoscopy and seems to be superior for detecting vitreoretinal traction associated with SBS. OCT even suggested small hemorrhagic macular retinoschisis in Patient 1 with perimacular folds.

Thus, OCT revealed and provided valuable additional information about the ocular lesions in these patients. Vitreoretinal membrane formation detected by OCT could support the pathophysiological theory of a direct mechanical effect. In infants, the vitreous is fully adherent to the posterior pole. In cases of extensive hemorrhagic retinal pigment epithelium (RPE)-detachments or preretinal blood accumulation, one may postulate certain forces pushing up the overlying vitreous body and consecutively causing traction and fold formation in the vicinity (Figure 5). In SBS, histopathologic examination has shown the retinoschisis being caused by blood dissecting between the outer nuclear layer and the inner segment of the retina.[19] But the retina may split apart at any of its layers with blood accumulating in the intervening cystic cavity. In Patient 1, cystic cavities were seen in the inner retinal layers and might have even affected the retinal nerve fiber layer.

Surprisingly florid neovascularization had developed in Patient 1 already three weeks after injury. Neovascularization of the optic disk following retinal schisis and hemorrhage from a shaking injury has been reported.[20] These authors suggested ischemia of superficial retinal layers attributable to retinoschisis and ischemia of the deeper retina due to disruption of the retinal capillary networks as possible mechanisms.

Optical coherence tomography examination is difficult in small infants and can not be performed routinely. In most cases, SBS can be diagnosed by regular fundus examination in combination with typical general findings such as subdural, and/or subarachnoidal hemorrhages, and encephalopathy. However, perimacular folds and hemorrhagic macular retinoschisis are regarded as highly specific for SBS and indicate poor visual outcome.[21,22] In our study, OCT revealed perimacular folds, retinal traction, and macular retinoschisis, which was not seen by ophthalmoscopy. Whether these OCT findings have the same specificity for diagnosis and prognosis is not clear yet. Further studies would be needed to test the clinical value of OCT in SBS. However, attributable to the difficult imaging conditions in these critically ill patients, it is unlikely that larger scale studies are possible, unless a hand-held OCT device would become available.

In conclusion, OCT revealed morphological changes missed by clinical examination that provided data in favor of the theory of vitreoretinal traction as a direct mechanical effect attributable to the rapid head movements in SBS.

THE AUTHORS INDICATE NO FINANCIAL SUPPORT OR FINANCIAL CONFLICT OF INTEREST. INVOLVED IN DESIGN AND conduct of study (V.S., M.M.); collection, management, analysis, and interpretation of the data (V.S., K.L., M.M.); and preparation and review of the manuscript (V.S., K.L., M.M.). Parents/legal guardians gave informed written consent to participate in the study, which was in compliance with Institutional Review Board regulations of the University of Zurich. The study adhered to the tenets of the Declaration of Helsinki and all federal and state laws.

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AJO History of Ophthalmology Series

William Osler, almost Ophthalmologist

When William Osler finished medical school at McGill University in 1872, he was interested in a career in ophthalmology, thinking it would give him the opportunity to pursue medical research in his leisure time. Howard, his mentor at McGill, encouraged him by saying McGill had no ophthalmologist and the post would be his if he could get training at Moorfields in London, England. Osler went to London and applied for the post of House Surgeon at Moorfields, but the post was given to Frank Buller instead. Buller, older than Osler, had already had training with von Graefe and Helmholtz, and was eventually to obtain the professorship at McGill and go on to a stellar career in Canada. Osler was already at work in the physiology laboratory of John Burden Sanderson at the University College Hospital, found the work congenial, gave up his plans for ophthalmology and eventually had his historic impact on general medicine and medical education. Nonetheless, his first paying patient when he went into practice, as recorded in his accounts ledger, was "removal of cinder from eye....50 cents."

Provided by Ronald Fishman, MD, of the Cogan Ophthalmic History Society.