

Dense Vitreous Hemorrhages Predict Poor Visual and Neurological Prognosis in Infants With Shaken Baby Syndrome

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ABSTRACT

Objective: A retrospective study was performed to examine the relationship between the severity of vitreous hemorrhages and the severity of neurological injury in infants with shaken baby syndrome, and the result of early vitrectomy in these infants.

Methods: Five infants, ages 3 to 8 months, with confirmed child abuse underwent ocular examination and neurological testing to identify varying degrees of vitreoretinal changes and neurological dysfunction. Pars plana vitrectomy was performed on one or both eyes in each infant to remove the vitreous hemorrhage.

Results: The three infants with bilateral dense vitreous hemorrhage and multiple subarachnoid hemorrhage and cerebral contusions had poor postoperative ocular and neurological outcomes including light perception (LP) or no light perception (NLP) vision, large retinal holes or tears, retinal ischemia, and severe encephalopathy. The other two infants presented with bilateral subhyaloid and retinal hemorrhages without dense vitreous involvement, along with mild subarachnoid hemorrhage and minimal neurological findings. Following vitrectomy, visual acuities in these infants improved to or remained at the fix and follow state, and both infants improved neurologically.

Conclusions: The presence of dense vitreous hemorrhage in infants with shaken baby syndrome has a poor visual prognosis, due to the frequent concomitant occur-

rence of significant retinal and visual cortical pathology. In contrast, infants with only intraretinal or subhyaloid hemorrhage tend to have a much better prognosis, with less disruption of both intraocular and intracranial structures.

INTRODUCTION

Caffey¹ first described battered babies with subdural hematomas, intraocular bleedings, and multiple long bone metaphyseal fractures. Years later, he called this syndrome the "whiplash shaken infant" syndrome or "shaken baby" syndrome, as shaking of infants was the probable major cause of such injuries.² This form of child abuse is usually characterized by minimal or absent external lesions, although the injury may be severe enough to cause intracranial hemorrhages or cerebral dysfunction leading to permanent neurological deficits.^{2,3} A high percentage (50% to 100%) of these infants develop intraretinal hemorrhages,^{4,7} and there have also been reports of macular scarring,⁸ retinal detachment,⁹ and optic atrophy.¹⁰ Wilkinson¹¹ et al have previously reported a significant correlation between severity of retinal hemorrhages and acute neurological findings in babies who have sustained whiplash/shaking injuries. In the present study, we have analyzed the relationship between severity of vitreous hemorrhages and severity of neurological injury in five cases of shaken baby syndrome, and further examined the result of early vitrectomy for vitreous hemorrhages in these infants.

MATERIALS AND METHODS

Our study includes five cases of shaken baby syndrome evaluated and confirmed as child abuse by the Social Work Services at the University Hospital between March 1993 and October 1994. Suspicion of shaking and/or child abuse and neglect was based on history, clinical findings including ocular examinations, and computed tomography (CT) or magnetic resonance imaging (MRI) scans of the brain. Histories were obtained from caretakers including physicians, social workers, and, in some cases, from law enforcement personnel. All the infants met the following criteria of the "shaken baby syndrome": evidence of intraocular hemorrhages with subdural or subarachnoid hemorrhages with interhemispheric blood as per CT or MRI scans; and

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history of suspected child abuse. All of these infants underwent a complete neurological examination along with CT or MRI scans of the brain and electroencephalogram (EEG) evaluation. Associated trauma data were obtained from physical examination, skull radiographs, and skeletal surveys. After initial evaluation by pediatricians and neurologists, all of these infants were referred to the ophthalmologist. Complete ocular examinations, including indirect ophthalmoscopy and B-scan ultrasounds, were performed on all of them.

These infants underwent pars plana vitrectomy for removal of vitreous hemorrhages under general endotracheal anesthesia. The vitrectomy involved a three-port technique with sclerostomy sites placed at about 2 mm posterior to the limbus.

RESULTS

The pre- and postoperative visual acuity, preoperative ocular findings, intraoperative findings, CT or MRI scan results, EEG or VEP results, and neurological course of these five infants are given in the Table. Pars plana vitrectomy was performed on these infants within a mean interval of 33 days (range 10 days to 2 months) between the time of presentation and time of surgery.

Patients 1, 2, and 3 had dense vitreous hemorrhages with acuities of light perception at the time of presentation. None of them improved visually after pars plana vitrectomy. All these infants with dense vitreous hemorrhages had other associated retinal pathologies including large retinal holes, macular holes, and retinal ischemia, as indicated by attenuated opaque retinal vessels. Patient 3 underwent a second vitrectomy for repair of a giant retinal tear. We found that these infants also had a poor neurological outcome as documented by the clinical course and EEG or VEP results. They all had severe encephalopathy or diffuse cerebral dysfunction.

Patient 1 had a subarachnoid hemorrhage involving the interpeduncular cistern and extension along the right tentorium cerebelli (Fig 1a). The patient also had a significant infarction involving the right occipital lobe and a frontal lobe contusion (Fig 1b). Subsequently, a bilateral frontal subdural fluid collection extending into the interhemispheric fissure developed which required drainage (Fig 1c). The EEG revealed a severe degree of encephalopathy and cerebral dysfunction. A flash VEP failed to reveal any response under any of the stimulus parameters. The brain-stem auditory evoked response (BAER) report indicated a possible hearing loss with impairment of brain stem function. He was hypertonic, agitated, and remained in the chronic vegetative state, without any response to the environment, and expired 5 months following the injury.

Patient 2 had a large right frontal, parietal, and temporal subdural hematoma with extension into the interhemispheric fissure. The child underwent right craniotomy with drainage of subdural hematoma. A follow-up CT scan demonstrated a right hemispheric infarction, most prominent in the right posterior cerebral artery distribution, requiring a right frontoparietal craniectomy, opening of

dura, and duraplasty with bovine dura. His left hemiparesis continued with multiple seizures due to increasing hydrocephalus for which he had an occipital shunt procedure. His EEG showed severe abnormalities in the right posterior hemisphere with higher voltage showing in the anterior temporal region of the left hemisphere (epileptic tendency).

Patient 3 had marked extensor spasticity and dystonic posturing. He developed generalized seizures with signs of devastating neurological deficit. A CT scan of the brain showed low attenuation of the cerebrum indicating diffuse cerebral ischemia as well as having a bilateral subdural hematoma (Fig 1d). His seizures progressed, and he developed hypoventilation with apneic spells. A follow-up CT scan 1 month later revealed marked cerebral atrophy (Fig 1e). His EEG showed severe, diffuse cerebral dysfunction, with frequent spike discharges from the anterior region, suggestive of an epileptic tendency.

Patients 4 and 5 had intraretinal and subhyaloid hemorrhages without dense vitreous involvement, and visual acuities of light perception (patient 4) and fixation and follow (patient 5) at the time of presentation. Postoperatively, the vision of both infants was fixation and follow. These infants did not have any other retinal complications, and the retinas remained attached. Patient 4 had a lensectomy for removal of cataract which secondarily developed after vitrectomy. We found that these infants without dense vitreous hemorrhages had a better neurological prognosis compared with those with dense vitreous hemorrhages. Both of these infants improved neurologically over the follow-up period.

Patient 4 initially had right frontal intracranial hemorrhages with interhemispheric subdural hematoma, and bilateral ischemia of temporal, parietal, and occipital lobes, with some edema. The early EEG revealed diffuse cerebral dysfunction with occasional sharp waves originating from both hemispheres consistent with an epileptic tendency. The seizures were controlled medically, and his neurologic status improved. Three weeks following his injury, a flash VEP was found to be normal for both eyes. A repeat EEG returned to normal activity without any evidence of epileptiform activity. The infant showed improvement in his neurological status.

Patient 5, at the time of initial presentation, had several episodes of apnea, cyanosis, and seizure activity, and a brain CT scan revealed bilateral frontal subdural hemorrhage (Fig 1f). A week later, the EEG was found to be normal in the wake state, without any epileptic activity, and the repeat brain CT scan revealed resolving hemorrhage within the subdural space (Fig 1g). The child improved neurologically during the hospital course and did not need any neurological intervention.

DISCUSSION

Our results show that the presence of dense vitreous and intracranial hemorrhage in infants with the shaken baby syndrome suggests a poor visual and neurological prognosis. In addition to the vitreous hemorrhages, all of these

TABLE
Summary of Patient Data

Patient no.	Age at onset	Duration*	Pre-/postoperative visual acuity	Preoperative ocular findings	Surgical procedure
1	7 mo	2 mo	LP/LP (OU)	Dense vitreous hemorrhages (OU); nystagmus (-); APD (+)	PPV (OU)
2	8 mo	10 days	LP/LP (OU)	Dense vitreous hemorrhages (OU); nystagmus (-); APD (-)	PPV (OU)
3	5 mo	1.5 mo	LP/LP (OD); LP/NLP (OS)	Dense vitreous hemorrhages (OU); nystagmus (-); APD (+)	1. PPV (OU) 2. Repeat PPV (OS)
4	3 mo	3 wk	F&F/F&F (OD) LP/F&F (OS)	Intraretinal hemorrhages (OD); subhyaloid macular hemorrhage (OS); nystagmus (-)	1. PPV (OS) 2. Repeat PPV (OS) 3. Lensectomy (OS)
5	4.5 mo	2 wk	F&F (OU)/ F&F (OU)	Subhyaloid and intraretinal hemorrhages (OU); nystagmus (-)	PPV (OD)

CT=computerized tomography; MRI=magnetic resonance imaging; EEG=electroencephalogram; VEP=visual evoked potential;

PPV=pars plana vitrectomy; LP=light perception; APD=afferent pupil defect; NLP=no light perception; F&F=fixes and follows;

*Time interval between onset of trauma and date of eye surgery.

infants had additional retinal complications, including large retinal holes, macular holes, and retinal ischemia. As a result of these complications, the visual outcome was poor despite the retinas remaining attached. These infants had severe encephalopathy and/or diffuse cerebral dysfunction and a devastating neurological course, and visual loss might be partly due to diffuse cortical damage. In contrast, those infants with intraretinal and/or subhyaloid hemorrhages had minimal, if any, retinal damage and had better visual and neurological results. A similar relationship between the severity of intraocular hemorrhages and the severity of acute neurological injury was previously shown in a study by Wilkinson et al,¹¹ although none of these infants underwent pars plana vitrectomy for removal of vitreous hemorrhages. To our knowledge, there have been no published reports evaluating the visual outcome in vitreous hemorrhages in infants with the shaken baby syndrome after pars plana vitrectomy.

It is very critical to determine a plan for vitrectomy intervention in infants with dense vitreous hemorrhage to prevent occlusion amblyopia. In children with congenital cataract, irreversible damage from occlusion amblyopia occurs as early as 6 weeks after monocular occlusion.^{12,13} It is not entirely clear how amblyopiogenic bilateral vitreous hemorrhage is, and how soon it needs to be evacuated to

prevent amblyopia. In a series of six infants with vitreous hemorrhage, Ferrone and deJuan¹⁴ recommend considering vitrectomy as early as 3 to 4 weeks after the onset of vitreous hemorrhage to avoid amblyopia. Apart from amblyopia, occlusion by vitreous hemorrhage may result in anisometropic myopia.^{14,15} In our series, vitrectomy was performed on the infants within 10 days to 60 days following the onset of vitreous hemorrhage. In spite of early intervention in infants with dense vitreous hemorrhages, the visual result was poor because of association of other retinal complications (ie, retinal holes and ischemia) as well as visual pathway dysfunction. In infants with subhyaloid and/or intraretinal hemorrhages, early vitrectomy resulted in acceptable visual recovery, probably due to the absence of other retinal or visual pathway complications, although it was uncertain for us to predict the visual outcome in these infants if the surgery was delayed.

The vitreous in the infant is more formed and gelatinous than in the adult. Eisner¹⁶ has demonstrated that the peripheral (cortical) zone of the vitreous body has the highest density and a greater resistance to mechanical stress than the more transparent, semi-fluid central vitreous. In infancy, there is very subtle differentiation between the peripheral and the central vitreous that becomes more pronounced with increasing age.¹⁷ This unique feature of the

Intraoperative findings	CT/MRI scan	EEG/VEP	Neurological course
Large, multiple retinal holes (OU); retinal ischemia (OU)	Occipital infarct; subarachnoid hemorrhage; frontal lobe contusion	Severe encephalopathy	Chronic vegetative state; no response to environment; did not improve over 5 months; expired at age 1
Large, multiple retinal holes (OD); superotemporal retinal hole (OS); ischemia (OU)	Right subdural hemorrhage (frontal, parietal, temporal)	Severe encephalopathy	Left hemiparesis; craniectomy with evacuation of blood; seizures; no neurological improvement
Macular holes (OU); Giant retinal tear (OS)	Bilateral subdural hemorrhage; Diffuse cerebral edema and ischemia; cerebral atrophy	Diffuse cerebral dysfunction	Marked extensor spasticity; Dystonic posturing; seizures; no neurological improvement
Retina attached; no holes (OS)	Right frontal intracranial hemorrhage with bilateral cerebral ischemia and edema; subdural hematoma	Early EEG: diffuse cerebral dysfunction with epileptic tendency; Late EEG: normal activity without epileptic activity; VEP: normal	Patient neurologically improved
Retina attached; no holes (OD)	Bilateral frontal subdural hemorrhage		Patient neurologically improved

vitreous gel in infants probably explains the delay in spontaneous absorption of significant vitreous hemorrhage seen in the shaken baby syndrome.

Several pathophysiological mechanisms have been described for intraocular hemorrhages in the shaken baby syndrome. First, intraocular bleeding may result from sudden rise in retinal venous pressure following an acute, rapid rise in intracranial pressure.^{7,18-20} Caffey³ suggested that infants have proportionately larger head sizes as well as poorly developed cervical muscles supporting the head during vigorous shaking of the chest, and are thus vulnerable to whiplash injuries. Second, rapid acceleration/deceleration of the vitreous associated with the violent shaking of the infant's head may result in trauma to the retina and retinal blood vessels, causing intraocular bleeding.²¹ Third, the transmission of subarachnoid hemorrhage, subdural hemorrhage, or increased intracranial pressure into the optic nerve sheath also may result in intraocular hemorrhages.^{18,22}

The pathophysiology of Terson's syndrome, a condition of vitreous hemorrhage in association with intracranial or subarachnoid hemorrhage, is similar to that of intraocular hemorrhage seen in the shaken baby syndrome. Schultz²³ et al postulates that an acute rise in intracranial pressure in Terson's syndrome is transmitted through the optic

nerve sheath distally, causing stretching of the intraorbital veins. This in turn raises the intraocular venous pressure, causing rupture of the intraretinal vessels and creating a vitreous hemorrhage.

Apart from intraretinal and vitreous hemorrhages, several other retinal changes have been reported in infants with the shaken baby syndrome. These changes include macular scarring, retinal detachment,⁹ retinoschisis,²⁴ and perimacular retinal folds.²⁵ In our series, two infants had multiple retinal holes and one had bilateral macular holes. Such an extent of retinal damage may be due to consequences of vitreous traction related to the severity of impact of the whiplash/shaking type injuries. Massicote et al²⁶ have described the partial detachment of the vitreous from the retina with remaining attachments to the internal limiting membrane at the apices of perimacular retinal folds and the vitreous base in infants with the shaken baby syndrome, implicating vitreous traction in the pathogenesis of perimacular retinal folds.

There was one infant who developed a giant retinal tear in the present study. Although it was uncertain whether such a serious complication was present at the time of the initial trauma, there was the possibility that it may have occurred during the initial vitrectomy. Prevention of a retinal dialysis or giant tear can be achieved by placing the

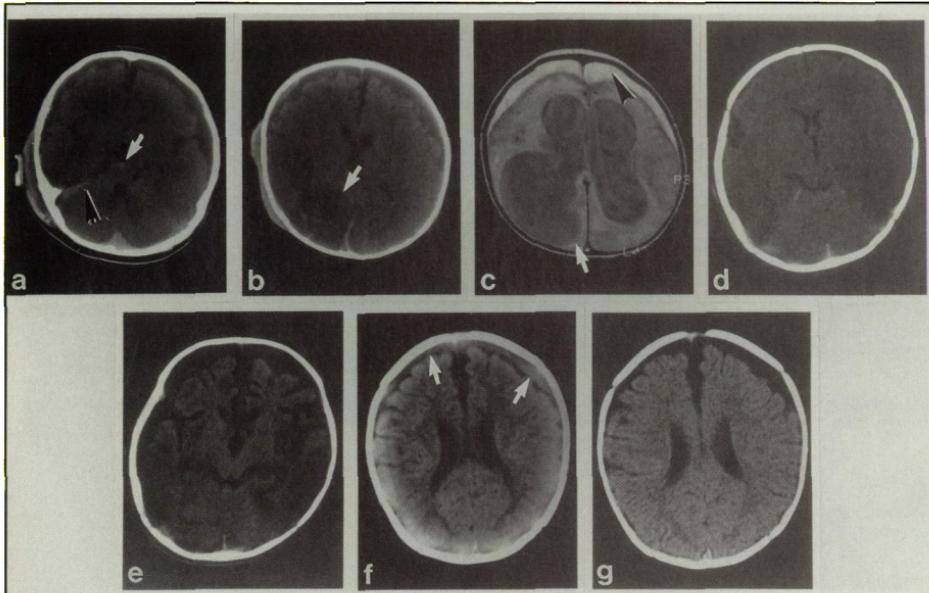


Fig 1: **a.** Brain CT scan of patient 1 illustrating a small subarachnoid hemorrhage within the interpeduncular cistern (arrow) and layering along the right tentorium cerebelli (arrowhead). **b.** Brain CT scan of the same patient showing decreased attenuation, representing infarction with punctate hemorrhages in the right occipital lobe (arrow) at the initial presentation. **c.** MRI scan 2 months later showing bilateral frontal subdural fluid collections (arrowhead). A focal compensatory dilatation of the right lateral ventricle extending into the area of the right occipital infarct (arrow) is

present. **d.** Brain CT scan of patient 3 illustrating decreased attenuation of the cerebrum relative to the cerebelli, supporting evidence for severe cerebral ischemia (CT reversal sign). **e.** Brain CT scan of the same patient one month later showing marked cerebral atrophy. **f.** Brain CT scan of patient 5 showing bilateral frontal subdural fluid collections including blood breakdown products (arrows). **g.** Brain CT of the same patient one week later showing reduced high attenuation indicating that the hemorrhage was resolving.

sclerotomies with a range of 0.5–1 mm posterior to the limbus rather than placing them at 2 mm posterior to the limbus as was performed in our study. Ferrone and deJuan¹⁴ have similarly reported retinal dialyses as a complication of vitrectomy in infants with vitreous hemorrhage because of placement of sclerotomies at 2 mm behind the limbus. Maguire and Trese²⁰ have suggested that sclerotomies placed at 0.5 from the limbus result in good success in two-port vitrectomies in infants.

Clinical signs seen in anterior visual pathway disease differ from those seen in posterior visual pathway disease. In anterior visual pathway dysfunction, the afferent pupillary defect and absence of nystagmus are usually present. For isolated posterior visual pathway lesions, pupils are usually spared, nystagmus may occur, and horizontal or vertical gaze paresis may present. The visual evoked potential is abnormal when anterior or posterior visual pathway injury is present.²¹ The electroretinogram is only useful for diffuse retinal pathology distal to the bipolar

neurons. In the present study, patients 1, 2, and 3 presented with clinical signs consistent with both anterior and posterior visual pathway involvement. In contrast, patients 4 and 5 had only media opacities and resolving intracranial hemorrhage without any significant retinal or visual pathway disruption. The absence of nystagmus in patients 1, 2, and 3 does not rule out the presence of posterior visual pathway involvement because the gaze centers might not be affected by the head injury.²² The poor visual outcome resulting in patients 1, 2, and 3 was due to a combination of both anterior and posterior visual pathway disruption, indicating that it is difficult to assign a relative proportion of each component responsible for the visual loss.

Dense vitreous hemorrhages in infants with the shaken baby syndrome indicate poor visual and neurological prognosis. Those infants with intraretinal hemorrhages or subhyaloid hemorrhages do better visually and neurologically. Although early vitrectomy may be indicated in infants with

dense vitreous hemorrhages to prevent occlusion amblyopia, the end result is still not encouraging because of association of other retinal and visual pathway complications.

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