Retinal Hemorrhage Predicts Neurologic Injury in the Shaken Baby Syndrome

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• **Shaken baby syndrome consists of intracranial and intraocular hemorrhage in the absence of signs of direct trauma in infants who have sustained whiplash/shaking injuries. We evaluated 14 consecutive cases of presumed shaken baby syndrome seen at the University of Michigan Hospitals, Ann Arbor, between August 1983 and March 1988, to determine if the severity of retinal hemorrhage was predictive of the severity of acute neurologic injury. The severity of retinal hemorrhage was based on the type and size of hemorrhage and the extent of fundus involvement. We found a significant correlation between retinal hemorrhage severity and acute neurologic findings. Diffuse fundus involvement, vitreous hemorrhage, or large subhyaloid hemorrhages were associated with more severe acute neurologic injury.**

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The shaken baby syndrome is a unique form of child abuse in which intracranial injury and intraocular hemorrhage may exist in the absence of external signs of direct head trauma.[1-7] In this syndrome, there is often a vague clinical presentation with minimal history of trauma, yet there may be substantial intracranial bleeding or other cerebral injury that may lead to permanent neurologic deficit and mental retardation.[1-3] Recently, Giangiacomo et al[4] reported retinal hemorrhages preceding the clinical and radiographic recognition of subdural hematoma in infants who have suffered whiplash/shaking-type injuries. The ophthalmologist may, therefore, be asked to evaluate an early manifestation of this condition before the complete evolution of its neurologic complications.

Although the association between intracranial injury and intraocular hemorrhage in battered or whiplash-shaken infants is well described, it is unknown whether a relationship exists between the severity of intracranial injury and severity of retinal hemorrhage. We attempted to determine whether such a relationship exists and to determine if certain clinically recognizable characteristics of retinal hemorrhage would help to identify infants at high risk for severe intracranial injury.

PATIENTS AND METHODS

The study consisted of 14 consecutive cases of shaken baby syndrome evaluated by the child protective services at the University of Michigan, Ann Arbor, between August 1983 and March 1988. Shaken baby syndrome was defined as findings of intraocular hemorrhage and intracranial injury in the absence of external signs of head trauma. During this period, all children suspected of being abused underwent a complete physical examination and radiologic studies as deemed necessary by the examining pediatrician. Children suspected of having intracranial injury underwent computed tomography of the head, and a fundus examination was performed by an ophthalmologist. When the children were in neurologically stable condition, their pupils were pharmacologically dilated to facilitate indirect ophthalmoscopy and fundus photographs. Ophthalmoscopy was performed and photographs were obtained within 72 hours of the injury.

An initial neurologic score was determined from the clinical examination and computed tomographic findings of the head obtained within the first week of presentation (Table 1). The scoring system was designed by a pediatric neurologist (M.D.R.) to reflect the hemorrhagic complications of the syndrome and the resulting clinical derangements as manifested in the neurologic examination. Two authors (M.D.R. and W.S.W.) who were "masked" to the fundus findings jointly determined the neurologic scores. A higher score reflected a more severe injury. A late neurologic score was assigned to 8 of the 12 patients who survived and were followed up for between 12 and 56 months. This score was assigned based on the presence of motor or cognitive delay, seizure activity, or residual focal deficit. Additionally, points were assigned for the presence of hydrocephalus or substance loss demonstrated on computed tomography of the head on follow-up examinations (Table 2). None of these characteristics was used in determining the initial neurologic scores.

The severity of retinal hemorrhage was judged from fundus photographs by one of us (D.P.H.) who was masked to the neurologic findings. The hemorrhagic retinopathy in all of our patients consisted of intraretinal or subhyaloid hemorrhages occurring primarily in a paravenous distribution, involving the peripapillary, macular, and peripheral regions of the fundus relatively independent of each other. Hemorrhage into the cortical vitreous was also seen. Classification of intraocular hemorrhage was based on clinically discernible characteristics (Table 3) relating to the type and extent of hemorrhage. Each eye was assigned a point score based on these characteristics, and each case was assigned a retinopathy score equal to that of the worst eye. In patients with equal retinopathy scores, eyes with larger subhyaloid hemorrhages or a larger number of intraretinal hemorrhages were ranked higher in

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severity. Spearman's Rank Correlation Test or, when appropriate, the independent t test was used to determine whether a correlation existed between the severity of retinopathy and the neurologic scores.

RESULTS

The mean age of the 14 patients (9 male and 5 female) when injured was 10.5 months (range, 2 to 28 months). Eleven of the 14 patients were younger than 1 year old. Ten patients were white, 3 were black, and 1 was of mixed racial descent. Two patients died within 48 hours of presentation. The frequency, laterality, range, mean, and median scores for the severity of intraocular hemorrhage are given in Table 4. The frequency, mean, and median of initial neurologic injury scores are given in Table 5. The initial and late neurologic injury scores according to the retinopathy score are given in Table 6. We found a significant correlation between the retinopathy score and the initial neurologic injury score ($N = 14$, $r = .574$, $.01 < P < .05$) and a corrrelation of borderline statistical significance between the retinopathy score and late neurologic injury score ($N = 8$, $r = .595$, $P > .05$). The relationship between a severe initial neurologic score (initial neurologic score greater than or equal to the median score of 6 points) and the type and extent of intraocular hemorrhage sustained is given in Table 7. Patients with vitreous hemorrhage, subhyaloid hemorrhage ($\geq$2 disc areas), or diffuse involvement (hemorrhage in all three fundus regions) had a significantly higher risk of severe acute neurologic injury ($P = .01$, Fisher's Exact Test).

Children with bilateral retinal hemorrhage tended to have more severe acute neurologic injury. Seven of 11 patients with bilateral retinal hemorrhages had initial neurologic scores of 6 or greater, whereas only 1 of 3 patients with unilateral retinal hemorrhages demonstrated a similar score. This difference was not statistically significant, however.

The mean age ($\pm$SD) of children with severe-type retinopathy was 6.4 ± 3.7 months compared with 14.6 ± 9.8 months for children without severe-type hemorrhage. This age difference was of borderline significance ($P = .058$, independent t test). A weak but nonsignificant correlation was found between patient age and retinopathy scores using Spearman's Rank Correlation Test ($r = .501$, $P > .05$). We found a strong correlation between the initial and late neurologic injury scores in the eight patients who were followed up for 12 months or more

Table 1.—Initial Neurologic Score*

Score Factors	Points
Level of consciousness	
Awake, alert	0
Somnolent	1
Obtunded, stuporous	2
Coma	3
Clinical neurologic findings	
Focal or generalized weakness	1
Seizure activity	1
Head circumference > 90th percentile or bulging fontanelle	1
Posturing	1
Head CT findings	
Subarachnoid hemorrhage	1
Subdural hematoma	1
Cerebral edema	1
Bilaterality of any of above CT findings	1

*Initial neurologic score equals the sum of point scores corresponding to presence of above characteristics (total points possible, 11). CT indicates computed tomographic.

Table 2.—Late Neurologic Score*

Findings	Points
Clinical	
Motor delay	1
Cognitive delay	1
Seizure activity	1
Residual focal deficit	1
CT†	
Hydrocephalus	1
Substance loss	1

*Late neurologic score equals the sum of point scores corresponding to presence of above clinical characteristics (total points possible, 6). CT indicates computed tomographic.

Table 3.—Classification of Intraocular Hemorrhage*

Hemorrhage	Points
Type (only one category chosen)	
Mild: Intraretinal hemorrhages only	1
Moderate: Subhyaloid hemorrhage, all lesions <2 disc areas in size	2
Severe: Subhyaloid hemorrhage, any lesion >2 disc areas in size, or vitreous hemorrhage	3
Extent (sum of categories below)	
Any hemorrhage within the following areas:	
Macula (>1 DD from disc, within 2 DD of fovea)	1
Peripapillary (within 2 DD of disc margin, excluding macula)	1
Periphery (outside macula or peripapillary regions)	1

*Retinopathy score equals the sum of scores from above two categories (total points possible, 6). DD indicates disc diameter.

Table 4.—Frequency and Laterality of Retinopathy Scores in 14 Patients With the Shaken Baby Syndrome*

Point Score	No. of Cases	Unilateral	Bilateral
0	0	0	0
2-3	5	1	4
4-5	4	2	2
6	5	0	5
Total Cases	**14**	**3**	**11**

*Mean retinopathy score: 4.4 points; median retinopathy score: 4 points.

Table 5.—Frequency of Initial Neurologic Injury Scores in 14 Shaken Infants*

Point Score	No. of Cases
0-3	3
4-6	7
7-9	4

*Mean initial neurologic injury score: 5.36 points; median initial neurologic injury score: 5 points.

Table 6.—Initial and Late Neurologic Injury Scores According to Retinopathy Scores

	Initial Neurologic Score		Late Neurologic Score	
Retinopathy Score	No. of Cases	Mean Score (Range)	No. of Cases	Mean Score (Range)
2	1	3 (3)	1	0 (0)
3	4	4.7 (1-9)	2	1 (0-2)
4	3	4.7 (2-8)	2	3 (2-4)
5	1	6 (6)	1	3 (3)
6	5	6.6 (5-9)	2	3 (3)
Total No. of Cases	**N = 14**		**n = 8**	
Spearman's rank correlation	$r = .574$, $P < .05$		$r = .595$, $P > .05$	

Table 7.—Relationship Between Initial Neurologic Score and Severity of Retinal Hemorrhage

Initial Neurologic Score	Presence of Severe Retinal Hemorrhage*	
	No	Yes
<6	6	0
≥6	2	6

*Defined as diffuse retinal hemorrhage (involving macular, peripapillary, and peripheral retina), subhyaloid hemorrhage 2 disc areas or larger in size, or vitreous hemorrhage. Fisher's Exact Test: $P = .01$.

($r = .976$, $P < .01$, Spearman's Rank Correlation Test).

COMMENT

Our results indicate that the severity of intraocular hemorrhage correlates with the severity of acute neurologic injury. The presence of subhyaloid hemorrhage greater than 2 disc areas in size, vitreous hemorrhage, or diffuse hemorrhage involving all three fundus regions (peripapillary, macular, and peripheral retina) was associated with a high initial neurologic score. In these children, the likelihood of severe neurologic injury seems to be much higher than in those without these characteristics. Although no statistically significant correlation was found between the retinopathy score and the late neurologic findings, there was a tendency for patients with high retinopathy scores to have more residual neurologic sequelae. Furthermore, children with severe initial neurologic injury were more likely to have more severe late neurologic sequelae. These findings suggest that an indirect relationship may exist between retinopathy, a predictor of acute neurologic injury, and late neurologic sequelae. It is possible that a larger number of patients with long-term follow-up would have provided statistical significance to this relationship. Nevertheless, the limited number of patients with long-term follow-up (two deaths, four patients unavailable for follow-up) precluded an adequate assessment of whether an association existed between retinopathy and late neurologic sequelae.

Although our data failed to demonstrate a statistically significant correlation between severe retinopathy and age, younger patients tended to have higher retinopathy scores. Many different pathophysiologic mechanisms have been proposed to explain the intraocular hemorrhages seen in the shaken baby syndrome and their relationship to age.[2,5,8-11] The tendency for more severe retinopathy in younger patients could be explained by clinical and experimental evidence that suggests that intraocular hemorrhage may occur from abrupt elevation in retinal venous pressure secondary to an acute, rapid rise in intracranial pressure.[10,12-14] Younger children weigh less, have greater head size in relation to body size, and have less developed neck musculature to support the head during shaking of the thorax. These factors may allow the heads of younger children to sustain a greater whiplash effect and more severe elevations in retinal venous pressure.[8]

In many cases of child abuse, the history is vague, without a clear-cut episode of shaking elicited. We cannot eliminate the possibility that the children in this study were subjected to more direct head trauma than indicated by the clinical examination and that the ocular manifestations we observed were a result of "battering" rather than a purely whiplash/shaking-type of injury. Nevertheless, our patient population seems to reflect an indirect mechanism of injury that is more consistent with whiplash/shaking based on the absence of clinically detectable signs of direct head trauma.

CONCLUSION

We found that the severity of retinal hemorrhage was predictive of the severity of acute neurologic injury in 14 children with the shaken baby syndrome. We also found the presence of large subhyaloid hemorrhages, vitreous hemorrhage, or diffuse involvement of the fundus to be useful in identifying battered or shaken children who are likely to have severe neurologic injury.

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