Long-term visual outcomes following abusive head trauma with retinal hemorrhage

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PURPOSE To report the rates of vision loss and select ocular findings after abusive head trauma (AHT) with retinal hemorrhages at a single center.

METHODS The study cohort was identified by review of billing records for patients presenting simultaneously with retinal hemorrhages and abusive head trauma at the Children's Hospital of Colorado from October 2005 to April 2017. The following data were analyzed: retinal examination at initial admission and visual acuity, other pertinent eye findings, and eye management at follow-up visits. Patients with <1 month of follow-up were excluded.

RESULTS Of 96 children, at last follow-up 46% had abnormal vision for the given age in at least one eye. Ocular findings included strabismus (43%), amblyopia (40%), optic disk pallor (13%), and cortical visual impairment (19%). For the 41 patients with strabismus, 20 (49%) required eye muscle surgery. Cortical visual impairment was almost three times higher in patients with strabismus compared with patients without strabismus ($P = 0.023$) and almost 6 times higher in patients with optic disk pallor than in those without ($P < 0.001$). Three patients (3%) required retinal surgery.

CONCLUSIONS In our study cohort, there was a high rate of long-term vision impairment and ophthalmologic comorbidities in children with AHT and retinal hemorrhage. (J AAPOS 2019;23:329.e1-4)

Abusive head trauma (AHT), also known as shaken baby syndrome, is the leading cause of child abuse deaths in the United States.[1] AHT is characterized by intracranial hemorrhages, long bone fractures, and retina hemorrhages in all layers of the retina. The retinal findings likely result from the unique shearing forces at the vitreoretinal surface that are created with rapid acceleration-deceleration of the head during shaking.[2] In infants, the vitreous is more adherent to the vitreous base and overlying vasculature, making tearing and hemorrhage much more likely when exposed to these shearing forces.[3] There are additional ocular findings that often present concurrently in these complex patients, including but not limited to macular scarring and retinoschisis, traumatic optic neuropathy, and cortical visual impairment (CVI).[4]

Although previous studies have shown that as many as half of survivors experience some visual impairment,[5-7] research on the prevalence of specific ocular findings and the long-term visual outcomes of children with AHT is limited. Our review of the literature found 2 case series that retrospectively analyzed long-term visual outcomes in AHT, but both studies were limited by the small sample size and lack of long-term follow-up.[8,9] The paucity of information makes it difficult to advise caretakers on expectations regarding visual recovery following after AHT. The aim of this study was to formally evaluate visual outcomes and associated ocular findings in children diagnosed with AHT with retinal hemorrhage at a single tertiary care referral facility.

Subjects and Methods

This study was approved by the Colorado Multiple Institutional Review Board. Billing records at Children's Hospital of Colorado were queried for patients presenting simultaneously with retinal hemorrhages (ICD 9 362.81, ICD 10 H35.60) and abusive head trauma (ICD 9 995.55, ICD 10 T74.4) from October 2005 to April 2017. At our institution, the diagnosis of AHT is made exclusively by the multidisciplinary Child Protection Team based on three criteria: (1) suggestive clinical and social history, (2) corroborating exam findings, and (3) a formal ruling-out process of mimicking disease. Retinal hemorrhage was selected as an inclusion criteria on the basis that this clinical finding is highly suggestive of

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shaking as the mechanism of injury as opposed to blunt force or single-impact trauma.[10]

Patients with follow-up of <1 month were excluded. Previous studies have shown that all intraretinal hemorrhage and most preretinal hemorrhage will clear rapidly within this time frame.[11] This minimum follow-up requirement allows for clearance of hemorrhages in order to assess accurately and begin to prognosticate long-term visual acuity.

Demographic data collected included sex, race, ethnicity, and date of birth. Examination date, location of retinal hemorrhages, presence or absence of retinal pigmentation, disk edema, and disk pallor were extracted from the initial encounter visit. Data abstracted from follow-up encounters at least 1 month after initial visit included examination date, visual acuity, presence of CVI, strabismus, amblyopia, and eye management including glasses, patching, atropine, strabismus surgery, and retina surgery. Patients were deemed to have strabismus or amblyopia based on either history during recovery or presentation at last follow-up.

The method of visual acuity testing was determined by the provider at the time of evaluation and was contingent on age and degree of cooperation. Methods of visual acuity measurement included fixation preference by induced tropia test, Teller Acuity Cards, Allen pictures, HOTV, and Snellen letters. Amblyopia was defined as a 2-line difference via Teller, Allen, HOTV, and Snellen, or fixation preference via induced tropia test. Normal visual acuity was defined as central-steady-maintained vision in both eyes in children <4 years of age. For HOTV and Allen testing, visual acuity norms for monocular testing were followed according to standards obtained from the Pediatric Eye Disease Study Group, with a threshold visual acuity of 20/63 in children 30 months of age, 20/50 for 3-year-olds, 20/40 for 4-year-olds, and 20/32 for 5-year-olds.[12] Teller visual acuity was defined as normal based on normograms established by Courage and colleagues,[13] with 95% prediction limits. Strabismus was defined as any ocular misalignment. The diagnosis of CVI was given to patients whose vision impairment was not fully explainable by their ocular findings and deemed to be due to neurologic injury.

Descriptive statistics included basic frequencies for categorical variables and means, standard deviations, and medians for continuous variables. Comorbidity rates of cortical visual impairment with select ocular comorbidities were compared with χ^2 testing for categorical variables. In addition to analysis of the cohort as a whole, patients were also stratified into subgroups based on the length of follow-up: 1 month to <1 year, 1 year to <3 years, 3 years to <5 years, and ≥5 years. Statistical significance was defined as a P value of <0.05.

Results

Of the 132 patients with AHT identified by search, 36 had follow-up of <1 month and were excluded, leaving a final study cohort of 96 children (72 males [75%]) with a diagnosis of both AHT and retinal hemorrhage. Mean patient age at first examination, during initial hospitalization for AHT, was 8.6 ± 9.1 months (standard deviation; range, 0.9-44.2). See Table 1. Patients were followed for an average of 30.3 ± 30.7 months (range, 1-145).

Table 1. Demographic characteristics of patients with abusive head trauma at Children's Hospital of Colorado

Patient characteristics	Result
Total patients	96
Sex, no. (%)	
Female	24 (25.0)
Male	72 (75.0)
Race/ethnicity, no. (%)	
White	52 (54.2)
Hispanic	21 (21.9)
African American	9 (9.4)
American Indian	2 (2.1)
Multiple race/ethnicity	6 (6.2)
Unknown	6 (6.2)
Age, months, at first exam	
Mean ± SD	8.6 ± 9.1
Median	5.3
Range	0.5-44.2
Follow-up, months	
Mean ± SD	30.3 ± 30.7
Median	20.5
Range	1-145

SD, standard deviation.

At final follow-up, 44 children (46%) were found to have abnormal vision for age in at least 1 eye. Other ophthalmologic findings included strabismus (41 [43%]), amblyopia (38 [40%]), and CVI (18 [19%]). See Table 2. Of the 41 children with strabismus, exotropia was most common (28 [68%]), followed by esotropia (12 [29%]) and hypertropia (1 [2%]). Twenty strabismic patients (49%) required eye muscle surgery. Of all patients, 36 (38%) received spectacle correction. Of the 38 patients with amblyopia, 33 (87%) were treated at some point during their clinical course with patching and 18 (47%) with atropine. Of all patients, 3 (3%) required retinal surgery: 2 required pars plana vitrectomy for nonclearing vitreous hemorrhage, and 1 required laser retinopexy for an operculated retinal hole. When analyzed by follow-up period, the rate of CVI, strabismus, amblyopia, and intervention generally increased for the groups with longer follow-up time (Table 2).

At initial examination, optic disk edema was present in 13 children (14%), with 3 unilateral cases and 10 bilateral cases. Only 1 of the unilateral cases required craniotomy for elevated intracranial pressure (ICP). The other 2 patients did not have elevated ICP and were monitored by Neurosurgery. Of the bilateral cases, 8 had elevated ICP requiring external shunt surgery (4), internal shunt surgery (3), or craniotomy (1) by neurosurgery. At final follow-up examination, optic disk pallor was present in 12 patients (13%) and retinal scarring in 17 patients (18%). See Table 3.

The comorbidity of CVI was almost three times higher in patients with strabismus than in patients without strabismus ($P = 0.023$) and almost six times higher in patients with optic disk pallor than in those without ($P < 0.001$). See Table 4. Children with amblyopia and disk edema were not

Table 2. Main ocular outcomes of children with abusive head trauma, stratified by follow-up time after initial visit

		Follow-up, no. (%)			
	Entire cohort, no. (%)	1 month to <1 year	1 year to <3 years	3 years to <5 years	5+ years
No. patients	96	33	34	17	12
Cortical visual impairment					
Yes	18 (18.8)	3 (9.1)	8 (23.5)	3 (17.6)	4 (33.3)
No	78 (81.2)	30 (90.9)	26 (76.5)	14 (82.4)	8 (66.7)
Strabismus					
Yes	41 (42.7)	7 (21.2)	15 (44.1)	11 (64.7)	8 (66.7)
No	55 (57.3)	26 (78.8)	19 (55.9)	6 (35.3)	4 (33.3)
Type of strabismus (n = 41)					
Esotropia	12 (29.3)	0 (0)	4 (26.7)	5 (45.4)	3 (37.5)
Exotropia	28 (68.3)	7 (100)	11 (73.3)	5 (45.4)	5 (62.5)
Hypertropia	1 (2.4)	0 (0)	0 (0)	1 (9.1)	0 (0)
Ambylopia					
Yes	38 (39.6)	6 (18.2)	13 (38.2)	11 (64.7)	8 (66.7)
No	58 (60.4)	27 (81.8)	21 (61.8)	6 (35.3)	4 (33.3)
Management					
Glasses	36 (37.5)	4 (12.1)	11 (32.4)	12 (70.6)	9 (75.0)
Patching	33 (34.4)	7 (21.2)	10 (29.4)	12 (70.6)	4 (33.3)
Atropine	18 (18.8)	3 (9.1)	5 (14.7)	8 (47.1)	2 (16.7)
Strabismus surgery	20 (20.8)	1 (3.0)	5 (14.7)	8 (47.1)	6 (50.0)
Retina surgery	3 (3.1)	1 (3.0)	1 (2.9)	0 (0)	1 (8.3)
Normal vision for age					
Yes	52 (54.2)	23 (69.7)	19 (55.9)	9 (52.9)	1 (8.3)
No	44 (45.8)	10 (30.3)	15 (44.1)	8 (47.1)	11 (91.7)

Table 3. Ocular characteristics in patients with abusive head trauma at Children's Hospital of Colorado

Retinal findings	No. (%)
Retinal scarring	
Yes	17 (17.7)
No	79 (82.3)
Location (n = 15)	
Fovea	7 (46.7)
Macula	8 (53.3)
Both	0 (0)
Nerve findings	
Disk edema	
Yes	13 (13.5)
No	83 (86.5)
Disk pallor	
Yes	12 (12.5)
No	84 (87.5)

Table 4. Presence of CVI by strabismus and amblyopia

Condition	CVI, no. (%)	No CVI, no. (%)	*P* value
Strabismus			
Yes	12 (29.3)	29 (70.7)	
No	6 (10.9)	49 (89.1)	0.023
Amblyopia			
Yes	8 (21.0)	30 (79.0)	
No	10 (17.2)	48 (82.8)	0.64
Disk edema			
Yes	2 (15.4)	11 (84.6)	
No	16 (19.3)	67 (80.7)	0.738
Disk pallor			
Yes	8 (66.7)	4 (33.3)	
No	10 (11.9)	74 (88.1)	<0.001

found to have significantly different comorbidity rates of CVI compared with children without the condition ($P = 0.640$ and $P = 0.738$, resp.).

Discussion

In general, AHT has the potential to cause lifelong multisystem injuries, instigating costly downstream effects. Ophthalmologic comorbidities are seen at a high rate in children with history of AHT. Our data shows that vision impairment is common, with almost half of children having abnormal best-corrected visual acuity for age in at least 1 eye at their final follow-up. The rate of strabismus was much higher in our cohort (43%) compared with the reported rate in the general population of 2%-3%.[14] This is not unexpected, because head trauma is known to provoke issues with motility and fusion. Similarly, amblyopia was more prevalent (40%) in our cohort compared with the rate of 2%-4% in the general population.[15] These high rates of amblyopia and strabismus are associated with a high rate of intervention (Table 2), specifically strabismus surgery (21%).

CVI occurred in approximately 1 in 5 children in our cohort, which is comparable to a previously published case series.[9] CVI is commonly defined as a loss in visual function in the absence of damage to the anterior afferent visual pathways or ocular structures.[16] Unlike in adults, who typically have clearly defined visual field deficits following visual pathway injuries, long-term manifestations of CVI in children are more heterogeneous and difficult to characterize.[17] Visual dysfunction includes decreased visual acuity, visual field deficits, and impairments in cognitive visual processing. Given the complexity

of diagnosis, its prevalence may be underrepresented, especially in those with minimal follow-up. This study demonstrates that children with strabismus are three times more likely to have concurrent CVI compared with those who do not have strabismus, supporting prior evidence that CVI and strabismus often coincide.

Optic disk edema at initial presentation was more prevalent than expected, found in 13 children (14%). In all cases of disk edema, subdural hemorrhage was present. Neurosurgical intervention for elevated ICP was more common in children with bilateral edema, in 8 of 10 children (80%), compared with cases with unilateral edema, in 1 of 3 children (33%). In the remaining 4 children without neurosurgical intervention, it is possible that the optic disk swelling may have been due to focal optic nerve injury.

Our study has several limitations, including the number of patients lost to follow-up: 36 patients (27%) examined in the hospital with AHT and retinal hemorrhage did not return to establish care at our institution beyond 1 month. We found that children who were followed longer had higher rates of poor vision, comorbidities, and interventions, compared with patients who had a shorter follow-up. Longer follow-up can lead to higher rates of detection of clinically significant ocular findings and insidious pathology like CVI, but it is more likely that healthier patients not needing as much care were lost to follow-up earlier. Therefore, visual outcomes from our cohort likely represent the more severe cases of AHT.

Despite limitations, the outcomes from this study were comparable to those presented by Oke and colleagues (J AAPOS 2016;20:e21 [Abstract 076]). Their slightly lower rates of strabismus (30%) and amblyopia (23%) may be explained by differences in the patient cohort. Their study did not require retinal hemorrhage as inclusion criteria. This could theoretically include patients with single-impact injury and not necessarily due to an acceleration-deceleration mechanism. Our study cohort, by contrast, was more homogeneous, requiring both AHT and presence of retinal hemorrhage.

The visual outcomes of two smaller retrospective studies must also be noted. First, in their study of 30 patients, McCabe and colleagues[8] examined the rate of ocular and radiographic findings during hospitalization to prognosticate visual impairment and mortality in children with AHT and retinal hemorrhage. They found that nonreactive pupils and midline shift of brain structures were associated significantly with mortality. At follow-up, 16 (73%) had at least fix-and-follow visual acuity. They concluded that initial visual acuity at hospitalization did not predict final visual acuity. Most children, however, were too young for Snellen visual acuity testing, which is suggestive of a short follow-up duration.

The second case series of 68 AHT patients (without retinal hemorrhage as a criterion) had a minimum of 2 months' follow-up and found that half of the surviving patients maintained good vision,[9] which was defined as fix-and-follow visual acuity at distance or 20/40 with Snellen testing. One-fourth of the patients had poor vision, defined as visual acuity of 20/200, unsteady fixation, or light perception vision. Patients whose initial vision was light perception or better during hospitalization were much more likely to see well at their last visit.

In conclusion, although prediction of visual prognosis must be individualized, our data provides a starting point for providers to educate caregivers regarding the visual outcomes to be expected following AHT with retinal hemorrhage. Our results indicate markedly elevated rates of decreased vision and visual comorbidities after AHT, highlighting the need for close monitoring to detect and treat these potentially vision-threatening processes.

References

1. National Center on Shaken Baby Syndrome. Learn more. National Center on Shaken Baby Syndrome. 2019. https://dontshake.org/learn-more;. Accessed March 3, 2019.
2. Levin AV. Retinal hemorrhage in abusive head trauma. Pediatrics 2010;126:961-70.
3. Narang SK, Estrada C, Greenberg S, Lindberg D. Acceptance of Shaken Baby Syndrome and Abusive Head Trauma as medical diagnoses. J Pediatr 2016;177:273-8.
4. Breazzano MP, Unkrich KH, Barker-Griffith AE. Clinicopathological findings in abusive head trauma: analysis of 110 infant autopsy eyes. Am J Ophthalmol 2014;158:1146-1154.e2.
5. Chevignard MP, Lind K. Long-term outcome of abusive head trauma. Pediatr Radiol 2014;44:S548-58.
6. Nuño M, Ugiliweneza B, Zepeda V, et al. Long-term impact of abusive head trauma in young children. Child Abuse Negl 2018;85:39-46.
7. King WJ, MacKay M, Sirnick A, Canadian Shaken Baby Study Group. Shaken baby syndrome in Canada: clinical characteristics and outcomes of hospital cases. CMAJ 2003;168:155-9.
8. McCabe CF, Donahue SP. Prognostic indicators for vision and mortality in shaken baby syndrome. Arch Ophthalmol 2000;118:373-7.
9. Kivlin JD, Simons KB, Lazoritz S, Ruttum MS. Shaken baby syndrome. Ophthalmology 2000;107:1246-54.
10. Binenbaum G, Mirza-George N, Christian CW, Forbes BJ. Odds of abuse associated with retinal hemorrhages in children suspected of child abuse. J AAPOS 2009;13:268-72.
11. Binenbaum G, Chen W, Huang J, Ying GS, Forbes BJ. The natural history of retinal hemorrhage in pediatric head trauma. J AAPOS 2016;20:131-5.
12. Pan Y, Tarczy-Hornoch K, Cotter SA, et al. Multi-Ethnic Pediatric Eye Disease Study Group. Visual acuity norms in pre-school children: the Multi-Ethnic Pediatric Eye Disease Study. Optom Vis Sci 2009;86:607-12.
13. Courage ML, Adams RJ. Visual acuity assessment from birth to three years using the acuity card procedure: cross-sectional and longitudinal samples. Optom Vis Sci 1990;67:713-18.
14. Friedman DS, Repka MX, Katz J, et al. Prevalence of amblyopia and strabismus in white and African American children aged 6 through 71 months the Baltimore Pediatric Eye Disease Study. Ophthalmology 2009;116:2128-2134.e1-e2.
15. Aldebasi YH. Prevalence of amblyopia in primary school children in Qassim province, Kingdom of Saudi Arabia. Middle East Afr J Ophthalmol 2015;22:86-91.
16. Hoyt CS. Visual function in the brain-damaged child. Eye (Lond) 2003;17:369-84.
17. Dutton GN, McKillop EC, Saidkasimova S. Visual problems as a result of brain damage in children. Br J Ophthalmol 2006;90:932-3.