

# Retinal Findings in Young Children With Increased Intracranial Pressure From Nontraumatic Causes

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abstract

**OBJECTIVES:** Increased intracranial pressure (ICP) has been suggested in legal settings as an alternative cause of retinal hemorrhages (RHs) in young children who may have sustained abusive head trauma. We assessed the prevalence and characteristics of RHs in children with increased ICP.

**METHODS:** We conducted a prospective, multicenter study of children <4 years old with newly diagnosed increased ICP as determined by using direct measurement and/or clinical criteria. Infants who were premature, neonates, and suspected survivors of abusive head trauma were excluded on the basis of nonocular findings. Fundus examinations were performed; extent, number, and type of RH in each of 4 distinct retinal zones were recorded.

**RESULTS:** Fifty-six children (27 boys) were studied (mean age 15.4 months; range 1–43 months). All of the children had elevated ICP that required intervention. One child had papilledema. No child (0%; 95% confidence interval: 0%–6.4%) or eye (0%; 95% confidence interval: 0%–3.3%) was found to have an RH. Causes of increased ICP included hydrocephalus, intraventricular hemorrhage, congenital malformations, malfunctioning shunts, and the presence of intracranial space-occupying lesions.

**CONCLUSIONS:** Although acute increased ICP can present in children with a pattern of peripapillary superficial RHs in the presence of papilledema, our study supports the conclusion that RHs rarely occur in the absence of optic disc swelling and do not present beyond the peripapillary area in the entities we have studied.



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**WHAT'S KNOWN ON THIS SUBJECT:** Although the literature suggests that retinal hemorrhages (RHs) beyond the peripapillary region are rare in children with increased intracranial pressure, the formation of widespread RHs in such children is often contested in legal proceedings regarding alleged abusive head trauma.

**WHAT THIS STUDY ADDS:** Increased intracranial pressure due to the entities we studied is an unlikely isolated cause of RHs beyond the peripapillary and prepapillary hemorrhages sometimes seen with papilledema.

**To cite:** Shi A, Kulkarni A, Feldman KW, et al. Retinal Findings in Young Children With Increased Intracranial Pressure From Nontraumatic Causes. *Pediatrics*. 2019; 143(2):e20181182

A limited body of current published literature indicates that increased intracranial pressure (ICP) does not cause retinal hemorrhages (RHs) in children except for the peripapillary and prepapillary hemorrhages associated with papilledema.<sup>1,2</sup> Despite this evidence, increased ICP continues to be implicated in legal settings as an alternative cause of RHs by the defense when young children are thought to have been survivors of abusive head trauma (AHT).<sup>2</sup> Attorneys often extrapolate from older studies with adult subjects to support this claim,<sup>3,4</sup> misuse studies to suggest that increased ICP can frequently cause widespread RHs in children,<sup>2,5-7</sup> or cite what they believe to be an inadequate body of literature to the contrary in children. Because accurate diagnoses are of critical importance in evaluations of child abuse, a broad evidence base is essential for clinicians to appropriately interpret potential findings of RH in cases of increased ICP. Increasing the evidence base is essential for both the bedside and the courtroom. Our purpose in this study was to assess the prevalence and characteristics of RHs associated with increased ICP in children through a prospective investigation of the ocular findings in a sample of children with acute increased ICP from causes other than child abuse.

## METHODS

We conducted a prospective, multicenter study of fundus examinations in children <4 years of age with increased ICP from March 2008 to February 2017. The study was approved by the Wills Eye Hospital Institutional Review Board (the data collection center) and by the institutional review boards of each of the 9 participating sites (Table 1). Because sites were accepted for participation on a rolling basis, the length of recruitment at each site varied. Inclusion required that eligible patients with increased ICP

**TABLE 1** Patient Recruitment

| Site  | No. Children Enrolled |
|---|-----------------------|
| Seattle Children's Hospital                   | 16                    |
| Children's Hospital Colorado                  | 8                     |
| The Hospital for Sick Children                | 7                     |
| Gillette Children's Specialty Healthcare      | 6                     |
| Children's Hospital of Philadelphia           | 5                     |
| Cincinnati Children's Hospital Medical Center | 4                     |
| Helen DeVos Children's Hospital               | 4                     |
| Arkansas Children's Hospital                  | 3                     |
| Nationwide Children's Hospital                | 3                     |

were identified by an on-site investigator or contact person in the neurosurgical service, general pediatric service, or PICU within 24 hours of admission. Given that families were to be approached so early in their hospital stay and at a time when they may have been in crisis because of the acutely elevated ICP in their children, possible impending surgery, and/or a new diagnosis, the primary inpatient attending physician was responsible for clearing the patients to have their families be approached for participation in the study. Therefore, an eligible patient was only excluded when the attending physician felt that the family was not emotionally in a position to offer adequate consent. Some patients may have been missed because of a failure to identify potentially eligible patients or unavailability of study staff to obtain consent or conduct an ophthalmic examination during the designated critical periods defined in the study. Data were not collected on unenrolled patients. Given the study structure, we believe it unlikely that the study sample was biased but rather that it represented a random sampling of eligible patients who were able to be enrolled. Consent for the release of deidentified data was obtained for all enrolled children; no child was excluded because consent was denied.

Increased ICP was determined by using clinical criteria and/or direct

measurements of ICP. Children who were included on the basis of clinical criteria exhibited (1) an egress of cerebrospinal fluid (CSF) under pressure during a surgical intervention and/or brain imaging that revealed a space-occupying lesion (or multiple lesions), cerebral edema, and/or hydrocephalus and (2)  $\geq 1$  symptoms or signs indicative of elevated ICP, including headache, vomiting, cranial nerve VI palsy, lethargy and/or drowsiness, conjugate downward deviation ("sunsetting") of the eyes, tense fontanelle or splayed sutures, clinical indicators of a failed ventricular shunt, and characteristic neuroimaging and/or optic disc swelling. When possible, ICP was also measured by using CSF shunt manometry or an intraventricular monitor. A direct ICP measurement of  $\geq 25$  cm H<sub>2</sub>O was used as a quantitative criterion for inclusion. This level was chosen on the basis of clinical expertise because there was no definitively established threshold for abnormally high ICP in infants and toddlers at the onset of the study. Infants <1 year of age who were born before 36 weeks' gestation, neonates <4 weeks postpartum, and children who were diagnosed by a multidisciplinary team as being survivors of AHT independent of the eye findings were excluded.

Child age and sex, medical history, ophthalmologic examination findings, brain imaging findings, ICP measurements, and the duration of elevation for increased ICP (if available) were collected from the medical record, deidentified, and sent to the data collection center by using fax or e-mail. Consent was not required for the eye examination because it was believed at the participating sites to be routine care for children with increased ICP. Informed consent was obtained from at least 1 parent or guardian for all enrolled children who met the study criteria to allow for the disclosure of

the deidentified data from their ophthalmic and neurosurgical care providers to the data collection site. The consent discussion included the disclosure of the possibility of an investigation for AHT should retinal examination reveal findings that might be concerning for possible child abuse.

Ophthalmologists at each site performed retinal examinations by using indirect ophthalmoscopy through pathologically or pharmacologically dilated pupils on each child. When the pupils were not pathologically dilated, the decision to use mydriatic eye drops (1 drop each of cyclopentolate or tropicamide 1% and phenylephrine 2.5%) with or without proparacaine 0.5% was based on the medical team's assessment of each child's clinical state. If dilation was felt to be contraindicated, an attempt was made to examine the retina without dilation. Children for whom the retinal view was inadequate were excluded from the study. The extent, number, and type of RH in 4 distinct zones of the retina (A, peripapillary; B, posterior pole excluding the peripapillary zone; C, peripheral retina posterior to a circle through the vortex veins; and D, peripheral retina anterior to the vortex veins and extending to the ora serrata) were assessed by using a previously tested description system.<sup>8</sup>

Standard descriptive statistics were calculated. The primary outcome was the prevalence of RHs, which was calculated as the proportion of children and proportion of eyes with RHs among all examined children. The Wilson<sup>9</sup> method was used to calculate the 95% confidence intervals (CIs) for the prevalence of RHs.

## RESULTS

Fifty-six children were enrolled (Table 2). Their clinical and ICP measurement findings are

**TABLE 2** Subject Demographics of 56 Children With Raised ICP

|                      | No. Children |
|----------------------|--------------|
| Age, mo <sup>a</sup> |              |
| 1–5                  | 15           |
| 6–11                 | 12           |
| 12–23                | 13           |
| 24–35                | 10           |
| 36–43                | 6            |
| Sex                  |              |
| Male                 | 27           |
| Female               | 29           |

<sup>a</sup> The mean age for all children was 15.4 months.

summarized in Table 3. Three children had concurrent conditions, including hypertension (2 children) and coronavirus upper-respiratory infection (1 child), which were not considered to be potential risk factors

**TABLE 3** Clinical Findings of Increased ICP Among 56 Children With Raised ICP

|   | No. Children   |
|---|----------------|
| Direct measurement of ICP                       | 19             |
| CSF shunt manometry                             | 9              |
| Intraventricular monitor                        | 9              |
| Method unknown                                  | 1              |
| Clinical criteria                               | 53             |
| Vomiting  | 27             |
| Lethargy and/or drowsiness                      | 19             |
| Tense fontanelle or splayed sutures             | 18             |
| Headache  | 9              |
| Sunsetting of eyes                              | 9              |
| Papilledema                                     | 9              |
| Cranial nerve VI palsy                          | 1              |
| Other clinical signs of increased ICP           |                |
| CSF egress                                      | 8 <sup>a</sup> |
| Increased head circumference                    | 8 <sup>a</sup> |
| Irritability and/or fussing                     | 5              |
| Paresthesias and/or weakness of the extremities | 4 <sup>a</sup> |
| Decreased oral intake                           | 3              |
| Decreased gross motor skills                    | 1              |
| Progressive loss of balance                     | 1              |
| Head tilt                                       | 1              |
| Gaze abnormalities                              | 1              |
| Seizures  | 1              |
| Estimated length of time ICP elevated           |                |
| Hours   | 4              |
| Days  | 17             |
| Weeks   | 27             |
| Uncertain                                       | 8              |

Patients may have qualified for a diagnosis of increased ICP on the basis of both clinical criteria and/or direct measurement.

<sup>a</sup> In 3 patients, the specified clinical sign was noted to be the only indicator of increased ICP.

for RHs. In the 2 children with hypertension, no signs of hypertensive retinopathy (vascular changes or exudates) were noted on examination. Hypertension has not been reported as a cause of isolated RHs in young children. No child had a confounding condition, such as hyponatremia, hypernatremia, or coagulopathy. All 56 children met the clinical criteria for increased ICP defined above and were therefore eligible for inclusion in the study on the basis of clinical criteria alone. Reliable, direct ICP measurements ranging from 25 to 50 cm H<sub>2</sub>O were also obtained in 7 children, who had a mean ICP reading of 28.9 cm H<sub>2</sub>O and therefore met quantitative criteria for inclusion in addition to clinical criteria. Twelve children had unreliable ICP measurements that were made only after a neurosurgical release of CSF had already occurred, resulting in values that were assumed to be falsely reduced by an indeterminable amount because of the preceding release of CSF. Measurements were obtained by using CSF shunt manometry in 9 children and intraventricular ICP monitors (including external ventricular drainage devices) in 9 children. The method of ICP measurement was not recorded for 1 child. No child had an intraparenchymal or subarachnoid bolt. The estimated duration of ICP elevation in most children was on the scale of weeks (with a range from <12 hours to 6–8 months in 1 patient), but all children, including those with chronic or intermittently elevated ICP, had acutely elevated ICP at the time of their admission that required intervention.

The etiologies of elevated ICP are summarized in Table 4. We had no children who presented with increased ICP caused by trauma. Imaging findings revealed hydrocephalus or ventriculomegaly in 45 children and the presence of a mass, tumor, or cyst in 18 children.

**TABLE 4** Causes of Increased ICP

| Cause  | No. Children |
|--|--------------|
| Brain tumor  | 15           |
| Primary hydrocephalus <sup>a</sup>   | 15           |
| Shunt obstruction  | 12           |
| Secondary hydrocephalus  | 16           |
| Arteriovenous malformation <sup>b</sup>  | 2            |
| Chiari malformation  | 3            |
| Craniosynostosis   | 3            |
| Dandy-Walker malformation (Joubert syndrome)                                     | 1            |
| Intraventricular cyst  | 1            |
| Intraventricular hemorrhage <sup>b</sup>   | 2            |
| Previous meningococcal disease and/or encephalitis <sup>a</sup>                  | 2            |
| Subdural hygroma   | 1            |
| Cerebral edema (birth anoxia with previous subdural and subarachnoid hemorrhage) | 1            |

<sup>a</sup> One child had both bacterial meningitis and hydrocephalus.

<sup>b</sup> One child had intraventricular hemorrhage from an arteriovenous malformation.

Intraventricular hemorrhage was seen in 2 children. In 13 children,  $\geq 1$  congenital malformation (type 1 Chiari malformation, schizencephaly, encephalocele, cerebellar hypoplasia, hydranencephaly, Dandy-Walker variant, syrinx, syringomyelia, and myelomeningocele) were seen. When appropriate, distinctions were made between imaging findings and etiologies of elevated ICP; the etiologies noted in Table 4 reflect the cause of elevated ICP at the time of the child's presentation. For example, 2 children presented with hydrocephalus due to shunt obstruction; these shunts had been placed previously for hydrocephalus that was caused by an intracranial cyst. In these patients, the elevation in ICP was attributed to the shunt obstruction rather than the cysts, although the latter were still noted on imaging.

No child was found to have RHs in any zone of the retina. The prevalence was 0% in any child (95% CI: 0%–6.4%) and 0% in any eye (95% CI: 0%–3.3%). In 51 of 56 children, there was documentation of an adequate assessment of all 4 retinal zones, including the posterior pole (zones A and B) and peripheral retina (zones C and D). In 5 children, the assessment of zone D (the outermost peripheral retina) was suboptimal,

although there were no RHs in zones A, B, or C, which comprise the majority of the retina. Additionally, for 2 children, details of the examination (ie, the quality of retinal view) were not provided, although there were no RHs noted in the assessment of any of the 4 zones. Successful pharmacologic dilation of both eyes was achieved in all but 2 children, 1 of whom was dilated in the left eye only but received a bilateral fundus examination. In the other child, a good view of the posterior pole in both eyes was obtained without dilation. Although 9 children were noted to have papilledema (a clinical sign of increased ICP) by a nonophthalmologist, results from an evaluation conducted by an ophthalmologist revealed optic disc swelling in 1 child who also did not have any RH.

## DISCUSSION

RHs in young children are a significant and well-established indicator of potential AHT, resulting from repeated acceleration and deceleration with or without blunt head impact (also referred to as "shaken baby syndrome").<sup>10</sup> RHs are present in ~85% of survivors,<sup>11</sup> frequently bilateral,<sup>11,12</sup> and most often located in multiple layers of the

retina, involving any geographic area from the peripapillary region to the ora serrata. In most patients, RHs are too numerous to count and widespread in distribution.<sup>10,13</sup> Increased ICP is a theory frequently proffered in legal settings as an alternative cause of severe RHs, usually in suspected cases of AHT. However, to our knowledge, no study in children has demonstrated that elevated ICP causes RHs beyond those associated with papilledema in the prepapillary or peripapillary distribution,<sup>1,2</sup> except for situations in which hyperacute rises in ICP occur, such as fatal head crushing,<sup>14–16</sup> fatal motor vehicle crashes,<sup>17–19</sup> ruptured aneurysms,<sup>6,7,20</sup> or a fall of 11 m.<sup>21</sup> Although medical conditions such as leukemia, coagulopathy, or sepsis may also produce RHs,<sup>22</sup> they are typically less severe. All of these conditions are readily obvious on the basis of history, basic laboratory testing, or neuroradiology.<sup>10</sup> As such, these recognizable entities cause little if any confusion regarding a diagnosis of AHT.

None of our subjects had RHs. A view of the entire retina was obtained in 91% of children. A full view of the far retinal periphery (zone D) could not be obtained in 5 children. We believe it is unlikely that RHs would be present in this most anterior zone yet absent in the other 3 zones that comprise the majority of the retina, especially because RHs from increased ICP are typically found on or around the optic nerve in association with optic disc swelling.

Our findings are consistent with those previously reported. In a study of 17 infants with increased ICP due to obstructive hydrocephalus, cerebral edema, spontaneous intracranial hemorrhage, or extraaxial effusion, the authors concluded that increased ICP without intracranial hemorrhage did not appear to cause severe RHs.<sup>23</sup> In survivors of AHT, there was no correlation between increased ICP and RHs.<sup>11</sup> Of children with a shunt

malfunction and a corresponding increased ICP, ~14% demonstrate papilledema, but RHs have not been noted.<sup>24,25</sup> Although the findings in older literature may suggest an association of RHs with conditions that cause increased ICP,<sup>26</sup> many of these studies were conducted at a time before AHT was recognized as a possible etiology.

Recent publications have clearly demonstrated the pattern of RHs seen with isolated increased ICP, including idiopathic intracranial hypertension. Binenbaum et al<sup>1</sup> found a small percentage (16%) of RHs among 100 children with nontraumatic elevated ICP but only in the presence of optic disc swelling; all of these were splinter and/or superficial intraretinal peripapillary hemorrhages and were limited to the area on or around the optic disc. Differences in subject age and cause of increased ICP between Binenbaum et al's<sup>1</sup> study and ours may explain why we found only 1 case of papilledema and no papilledema-related RHs in our subjects. For example, the most frequent cause of increased ICP in their study was idiopathic intracranial hypertension (70% of patients), whereas we had no patients with this etiology in our study. The prevalence of RHs and papilledema also appears to be higher in older children with fused cranial sutures; the age range of children in our study was 1 to 43 months (mean 15.4 months) compared with 3 to 17 years (mean 12 years) in the previous study.

Minns et al<sup>5</sup> found RHs in 38% of children (24 of 63) with raised ICP (as determined by using direct measurement, radiologic signs, or clinical signs) from a variety of causes. In children with nontraumatic encephalopathy and raised ICP, the incidence of RHs was 27% (4 of 15 children); all subjects with RHs lacked specific ICP measurements (4 of 9 children) and specifications of the cause of their encephalopathy. All

children with AHT and increased ICP (14 of 14) and 4 of 7 children with AHT without increased ICP had RH (total 86%; 18 of 21 children). This compares with the children with unintentional traumatic brain injuries (TBIs); 18% of children (6 of 34) with increased ICP and 4% of children (1 of 23) without increased ICP had RH (total 12%; 7 of 57 children).

Insufficient data are given regarding the number or pattern of RHs in the children who were not abused, thus limiting interpretation of their results. Because the types and severities of trauma and the underlying diseases were not specified for the children with unintentional trauma and the children with nontraumatic encephalopathy and RHs, respectively, it cannot be determined if their underlying illnesses and injuries, such as motor vehicle crashes<sup>17</sup> or leukemia, could be expected to cause RHs. The authors found no significant association between ICP (as determined by using direct measurement only) and either the prevalence or number of RHs.

Although they did find a statistically significant association between raised ICP (as determined by using direct measurement or clinical and/or radiologic signs) and RHs, the authors note that they cannot establish causation. In addition, this association was based on an analysis of patients in which researchers did not distinguish between children with inflicted TBIs ( $n = 14$ ), unintentional TBIs ( $n = 34$ ), and nontraumatic encephalopathy ( $n = 15$ ). Because increased ICP is frequently seen in inflicted TBI, the mixing of these samples will skew the analysis. The largest number of RHs was found in patients with inflicted TBIs; the authors concluded that the RHs in these patients "occurred at the time of the traumatic acceleration/deceleration injury" and were unlikely to have been exacerbated by increased ICP.

Although studies in adults have suggested that intraocular and optic

nerve sheath hemorrhages can occur after acute increases in ICP,<sup>3,4,27</sup> these findings are not always generalizable to children.<sup>2,28</sup> Anatomic and physiologic differences between the eyes of adults and children, particularly in the retinal vasculature and the strength of vitreoretinal attachments, must be considered along with differences in the etiology and nature of the raised ICP. The authors of 1 study of increased ICP found postmortem optic nerve sheath hemorrhages in 87% of eyes (40 of 46) and intraocular hemorrhages in 37% of eyes (17 of 46). They concluded that these hemorrhages were the result of ruptured intradural and bridging vessels and obstructions of the central retinal vein and retinochoroidal anastomoses.<sup>3</sup> Patient ages were not specified, but the article comes from an institution in which only adults are treated. In addition, all of the systemic diagnoses involved significant cerebral compression (severe cerebral trauma, spontaneous intracerebral hemorrhage, ruptured berry aneurysm, and internal carotid occlusion resulting in cerebral swelling without hemorrhage). These diagnoses are far more common in adults and were not seen in any of our patients. In another study, subhyaloid hemorrhages after subarachnoid, subdural, and extradural hemorrhages were seen in 5 of 6 adults with repeated episodes of sudden increases in ICP<sup>4</sup>, which are circumstances that rarely if ever occur in children without a corresponding history.

Those who have claimed that more widespread hemorrhages can occur because of raised ICP base their hypotheses on a presumed obstruction of venous outflow in the central retinal vein and a resulting increase in central venous pressure. Central retinal vein occlusion presents with a distinct pattern of retinal bleeding along with dilated and tortuous retinal veins.<sup>29</sup> In

a study of 17 children with nontraumatic increased ICP, there were no patients with retinal venous dilation.<sup>23</sup> The anatomy of retinal vasculature should prevent the formation of RHs in that there are collateral pathways for venous drainage through the vortex veins and retinochoroidal anastomoses.<sup>2,3</sup> Additionally, RHs are not seen in many other conditions that cause increases in central venous pressure, such as severe cough, vomiting, seizures, or cardiopulmonary resuscitation with chest compressions.<sup>2,30-33</sup>

Our study had certain limitations. Enrollment over the 9-year study period was limited because of a failure to identify potentially eligible patients, determinations by the primary attending team that families were not appropriate to be approached for consent during the acute phases of their children's care, and a lack of on-site investigator availability either for recruitment from the neurosurgical service and/or PICU or regarding ability to perform ophthalmologic examinations before patient discharge, resulting in a small sample size. We believe the children included in this study are unlikely to represent a biased selection given the random nature of these exclusions. We also did not have any patients present with hyperacute sudden elevations in ICP. Although an approximate duration of increased ICP was obtained for most children, the length of time was unknown in 8 children; however, they had mechanisms that would not be expected to result in

a hyperacute ICP rise. In all others, estimates ranged from hours to months. In children whose ICP had been elevated for weeks or months, hyperacute increases could have perhaps occurred transiently, producing RHs that resolved before their admission and study enrollment. However, preretinal and subretinal hemorrhages often take several weeks to resolve, and if these types of hemorrhage had indeed occurred, they likely would have been seen on examination. In addition, all of our subjects had ICPs elevated enough to require admission and/or surgical intervention at the time of their eye examinations. The absence of papilledema in almost all of our subjects can also be used to argue that their increased-ICP onsets of concern were acute.<sup>34,35</sup> Increased ICP was diagnosed clinically in all subjects, although a minority of our subjects had direct ICP measurements, and of these, many were felt to be unreliable. Obtaining accurate ICP measurements was not always possible given the necessity of CSF drainage in many patients with acutely elevated ICP, particularly because higher ICP values induce substantial artifactual reductions in measured ICP at both rapid and reduced rates of drainage.<sup>36</sup> Intraoperative confirmation of increased ICP was often made visually by the investigator rather than by using objective measurement.

We did not include children with AHT who had increased ICP to avoid confounding etiologies of RH. To evaluate the potential of increased ICP as a cause of RH, a population of

children with isolated increased ICP should be studied. If increased ICP is indeed able to cause RHs beyond the peripapillary area, then it should be able to do so regardless of etiology.

## CONCLUSIONS

In our study of 56 children with increased ICP due to primary and secondary hydrocephalus, shunt obstruction, brain tumors, and cerebral edema, we found no cases of increased ICP causing RH regardless of the etiology of increased ICP, age of the child, or duration of ICP elevation and despite a clinical severity of increased ICP in all children that warranted intervention. Although acute increased ICP can present in children with a pattern of peripapillary superficial RH in the presence of papilledema,<sup>1</sup> with our results, we provide clear evidence to support the conclusion that RHs rarely occur in the absence of optic disc swelling and do not present beyond the peripapillary area in the entities we have studied.

## ACKNOWLEDGMENT

We thank Gui-Shuang Ying for his assistance in the statistical analysis of the data.

## ABBREVIATIONS

AHT: abusive head trauma  
CI: confidence interval  
CSF: cerebrospinal fluid  
ICP: intracranial pressure  
RH: retinal hemorrhage  
TBI: traumatic brain injury

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designed the study, coordinated and supervised data collection and analysis, drafted the initial manuscript, and reviewed and revised the manuscript; and all authors approved the final manuscript as submitted and agree to be accountable for all aspects of the work.

**DOI:** <https://doi.org/10.1542/peds.2018-1182>

Accepted for publication Nov 14, 2018

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**FINANCIAL DISCLOSURE:** The authors have indicated they have no financial relationships relevant to this article to disclose.

**FUNDING:** Funded in part by the Foerderer Fund (Dr Levin), the Robison D. Harley, MD, Endowed Chair in Pediatric Ophthalmology and Ocular Genetics (Dr Levin), and the National Eye Institute (grant K12 EY-015398; Dr Binenbaum). Funded by the National Institutes of Health (NIH).

**POTENTIAL CONFLICT OF INTEREST:** Drs Levin and Feldman have provided paid expert testimony in child abuse cases; the other authors have indicated they have no potential conflicts of interest to disclose.

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*Pediatrics* 2019;143;

DOI: 10.1542/peds.2018-1182 originally published online January 10, 2019;

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