

Ophthalmologic findings in infants after an apparent life-threatening event

R.L. ALTMAN¹, S. FORMAN², D.A. BRAND^{1,3,4}

¹Department of Pediatrics, New York Medical College and Westchester Medical Center, Valhalla

²Department of Ophthalmology, New York Medical College and Westchester Medical Center, Valhalla

³Department of Medicine, New York Medical College, Valhalla, NY

⁴Office of Health Outcomes Research, Winthrop University Hospital, Mineola, NY - USA

PURPOSE. *An apparent life-threatening event (ALTE) refers to the sudden occurrence in an infant of a breathing abnormality, color change, or alteration in muscle tone or mental status. The finding of retinal hemorrhage in an infant who has experienced an ALTE strongly suggests the possibility of shaken baby syndrome. Previous ALTE studies have reported the frequency of this specific ocular abnormality but have not reported other ocular findings and their possible diagnostic significance.*

METHODS. *This study reviews ophthalmologic findings from a series of 120 patients with ALTE who underwent eye examinations as part of their inpatient evaluation. Details of the history, physical examination, diagnostic testing, and discharge diagnosis were abstracted from the medical record while the patient was in the hospital.*

RESULTS. *Ten patients (8.3%) had positive findings, including retinal hemorrhages in four of the six patients with shaken baby syndrome, the only ophthalmologic finding that helped establish the cause of an ALTE.*

CONCLUSIONS. *Since the funduscopic examination can help identify child abuse as the cause of an apparent life-threatening event, ophthalmologists play a critical role in the early evaluation of infants who have experienced such an event. (Eur J Ophthalmol 2007; 17: 648-53)*

KEY WORDS. *Apnea, Apparent life-threatening event, Infant, Retinal hemorrhage, Shaken baby syndrome*

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INTRODUCTION

Shaken baby syndrome is a form of child abuse in which a small infant is shaken by an adult. The head rotates forcefully on the neck, producing subdural hemorrhages, generalized cerebral edema, and characteristic eye lesions: multilayered symmetric or asymmetric retinal hemorrhages and, less commonly, vitreous hemorrhage and retinoschisis. The neurologic and ophthalmologic findings associated with shaken baby syndrome are well-documented in the literature (1, 2). The death rate ranges from 12 to 27%, and two-thirds of survivors suffer permanent neurologic sequelae that range from mild learning disabilities to mental retar-

dation, epilepsy, blindness, paralysis, and coma (3). Investigators have recently discovered that victims of shaken baby syndrome may come to medical attention for the first time when a caregiver brings the child to the hospital following an apparent life-threatening event (ALTE) (4, 5). An ALTE refers to the sudden occurrence of symptoms that appear frightening to the observer, typically the infant's caretaker (6). The symptoms include some combination of breathing abnormality that suggests decreased aeration, color change, alteration in muscle tone or mental status, choking, or gagging. Believing that the infant may die, the observer often rushes the infant to a hospital or calls 911. While the baby usually recovers quickly and

suffers no lasting ill effects, these symptoms may signal a serious disorder.

Numerous diseases and disorders can precipitate ALTEs of varying severity (7-9). Respiratory infections, gastroesophageal reflux, and seizures are the leading causes. Sepsis, congenital heart disease, shaken baby syndrome, and cerebral artery infarct are examples of the many other etiologies that have been implicated. Infants who have experienced an ALTE typically undergo extensive testing in the hospital, but 16 to 39% of them are nevertheless discharged without a diagnosis (7-9).

In 2003, we reported the relative frequency of different causes of an ALTE based on a 32-month consecutive series of ALTE admissions to a children's hospital (7). Shaken baby syndrome proved to be the cause in 2.3% of cases. The present investigation, which reviews the ophthalmologic data from the above case series, considers the potential role of a complete eye examination in identifying various conditions associated with ALTEs. This report describes the ophthalmologic findings from that series and the implications concerning the need for ophthalmology consultation in the ALTE context.

METHODS

The Maria Fareri Children's Hospital at Westchester Medical Center is a 114-bed tertiary care referral hospital located 20 miles north of New York City in Westchester County. The Medical Center is the major academic teaching affiliate of New York Medical College. It serves patients representing a broad demographic and socioeconomic spectrum and a geographic region that includes urban, suburban, and rural areas in the Hudson Valley of New York State, northern New Jersey, western Connecticut, and New York City.

The present report is based on data from the larger study, referred to above, that included all infants under 12 months of age who were admitted to the Children's Hospital between November 1, 1996, and June 30, 1999, for evaluation of an ALTE (n=243) (7). To have been enrolled in the overall ALTE study, a patient must have experienced the sudden occurrence of one or more of the following symptoms (from the initial history) which frightened the caregiver enough to seek medical attention: breathing irregularity (e.g.,

apnea, labored or shallow breathing, choking, gagging); color change (e.g., cyanosis or pallor); or altered muscle tone or mental status (e.g., hypotonia, hypertonia, clonic movements, unresponsiveness). These enrollment criteria constituted our working definition of an ALTE. The criteria are consistent with the official definition established in 1987 by the National Institutes of Health Consensus Development Conference on Infantile Apnea and Home Monitoring (10). A physician data manager visited the pediatric floor of the hospital and the 14-bed pediatric intensive care unit on a daily basis. She identified patients meeting the above inclusion criteria by examining the charts of all newly admitted infants. She followed these patients until discharge, recording on a standardized data collection instrument detailed information including the initial history and physical examination, diagnostic testing done in the hospital, and the final diagnosis.

The present study reports the ocular findings in the group of patients who underwent a complete ophthalmologic examination by an attending ophthalmologist as part of their evaluation (n=120). The remaining patients did not undergo such an examination because the responsible attending did not request an ophthalmologic consult. This occurred primarily when the cause of the ALTE seemed immediately apparent and could be easily confirmed, making a detailed ophthalmologic evaluation appear to be unnecessary. A common example was a patient quickly found to have bronchiolitis-associated apnea.

Since this was an observational study, the investigators did not impose a diagnostic testing protocol or attempt to influence physicians' decisions about obtaining tests or consultations. If a patient was admitted for an ALTE more than once during the study period, only the first admission has been included. The study, including the above procedure for collecting data from medical records, was approved by the Committee for Protection of Human Subjects, the institutional review board of New York Medical College.

RESULTS

Table I presents the demographics of the 120 study patients, and Table II shows their final diagnoses and the results of their ocular examinations. A total of 23

TABLE I - PATIENT DEMOGRAPHICS (N=120)

	No. (%)
Sex	
Male	65 (54.2)
Female	55 (45.8)
Age, mo	
<2	65 (54.2)
2–5	42 (35.0)
≥6	13 (10.8)
Race/Ethnicity	
Black	18 (15.0)
White, Hispanic	8 (6.7)
White, other than Hispanic	49 (40.8)
Other or unknown	45 (37.5)

different diseases or disorders were responsible for the 120 ALTEs. The most common causes were gastroesophageal reflux (42 patients), bronchiolitis (9 patients), seizures (7 patients), and shaken baby syndrome (6 patients). The latter diagnosis required evidence of an intracranial hemorrhage that could not be explained by an accident or nontraumatic process and that was accompanied by other supportive findings consistent with child abuse. Those supportive findings included unexplained fractures involving ribs, long bones, or skull; unexplained abrasions or ecchymoses of the head, face, neck, or chest; and retinal hemorrhage or detachment (7). Ten of the 120 ophthalmologic examinations (8.3%) yielded positive findings, including 4 instances of retinal hemorrhage in patients with a final diagnosis of shaken baby syndrome. One other positive finding—optic atrophy—may have been related to the cause of the ALTE (infantile spasms), but it did not play a

TABLE II - RESULTS OF OCULAR EXAMINATIONS PERFORMED IN PATIENTS ADMITTED TO THE HOSPITAL AFTER AN APPARENT LIFE-THREATENING EVENT

Diagnostic category	All patients (n=120)	Patients with positive findings (n=10)	Findings	Diagnosis
Gastrointestinal causes*	43	2	Icteric sclerae; maturing retina	Gastroesophageal reflux
Neurologic causes†	18	5	Poor visual fixation for age	Gastroesophageal reflux
			Nerve fiber layer hemorrhages (bilaterally); retinal detachment	Shaken baby syndrome
			Nerve fiber layer hemorrhages (bilaterally)	Shaken baby syndrome
			Nerve fiber layer hemorrhages (bilaterally)	Shaken baby syndrome
			Extensive nerve fiber layer hemorrhages (bilaterally) and preretinal hemorrhage (left eye)	Shaken baby syndrome
Infectious causes‡	17	1	Optic atrophy (bilaterally)	Infantile spasms
			Immature retina	Sepsis
Other causes§	12	1	Congenital nasolacrimal duct obstruction	Inborn error of metabolism
Unknown cause	30	1	Slightly blunted macular reflex; delay in visual maturation	Unknown

When more than one patient had the same diagnosis, the number of patients is shown in parentheses below. When more than one discharge diagnosis was listed in a patient's medical record, the diagnosis most likely to have caused the apparent life-threatening event is reported here.

*Gastroesophageal reflux (42), upper gastrointestinal bleed.

†Seizure (7), shaken baby syndrome (6), apnea of prematurity (2), infantile spasms (2), cerebral artery infarct.

‡Bronchiolitis (9), pneumonia (4), bacterial meningitis (2), sepsis, urinary tract infection.

§Choking episode (2), breath holding spells (2), nasopharyngeal reflux, laryngomalacia, inborn error of metabolism, birth trauma, vasovagal response, dehydration, drug ingestion, feeding difficulty

role in establishing the diagnosis. The five remaining findings – icteric sclerae, immature retina, poor fixation, nasolacrimal duct obstruction, and blunted macular reflex – were associated with conditions unrelated to the cause of the ALTE. Retinal hemorrhage was the only positive finding that helped focus the diagnostic evaluation by suggesting an etiology of the ALTE, or helped confirm a diagnosis.

DISCUSSION

We are not aware of any previous study that has reported all ocular findings from a series of patients with ALTE who have undergone ophthalmologic examinations. The study by Pitetti and associates (5), which was designed to identify cases of child abuse by looking for retinal hemorrhages, did not report results of eye examinations other than the presence of this specific finding. The present study, which examines data from the largest series of eye examinations in patients with ALTE of which we are aware, reports all positive ophthalmologic findings and the possible diagnostic significance of those findings. These data reveal that no abnormality besides retinal hemorrhage contributed to establishing the cause of an ALTE. The eye examination provides critical information for the detection of abusive head injury (2, 11-13). Studies have reported ocular abnormalities in 60 to 95% of the victims (14). These abnormalities have included direct, overt blunt injury to the eyelids, conjunctiva, lens, vitreous, and anterior or posterior segments, as well as indirect injuries associated with shaken baby syndrome. In our study, four of the six patients with a diagnosis of shaken baby syndrome had retinal hemorrhages, findings that helped establish the cause of the ALTE.

Shaken baby syndrome can produce retinal hemorrhages by several possible mechanisms (15, 16). When a baby is held around the chest or abdomen and shaken forcefully, either intracranial hemorrhage or increased intrathoracic pressure can produce elevated intracranial pressure that can be transmitted to the retinal vein and chorioretinal anastomoses or to the venous sinuses, leading to acute venous stasis and hemorrhage. It is also postulated that shearing forces generated by cycles of rapid acceleration and deceleration of a shaken baby's head may, themselves, be

sufficient to produce retinal bleeding or detachment. While retinal hemorrhages are known to be associated with shaken baby syndrome, inflicted injury is not the only cause of such lesions. Between 30% and 50% of normal newborns exhibit some retinal hemorrhaging, which tends to occur more frequently among older mothers or with vacuum-assisted deliveries (17). Approximately 90% of these hemorrhages resolve by 2 weeks of age. Retinal hemorrhages have also been reported in patients who have had a seizure, but the incidence in this group is less than 1% (18-20). Although the literature contains numerous case reports of retinal hemorrhages after cardiopulmonary resuscitation (21-23), the cause-effect relationship remains controversial: very few case reports have documented the absence of hemorrhages before resuscitation. Other possible causes of retinal hemorrhages include leukemic leukostasis, bleeding dyscrasias, bacterial meningitis, subacute bacterial endocarditis, central nervous system malformations, profound hypernatremic or hyponatremic dehydration, subarachnoid hemorrhages, or accidental injury (24-28).

A potentially catastrophic feature of shaken baby syndrome is that an infant who has been shaken may sustain neurologic injury but appear well with no external signs of abuse upon presentation for clinical evaluation (4, 29). The younger the victim, the more likely the physician will misdiagnose this form of child abuse (29), an error that can have dire consequences. Since shaken baby syndrome is diagnosed in 2 to 3% of patients admitted to the hospital after an ALTE (5, 7), and since up to 39% of patients with ALTE are discharged without a diagnosis (8), it is essential for physicians caring for infants after an ALTE to consider the possibility of occult inflicted injury.

In our ALTE series, when the cause of the ALTE was evident soon after a patient's initial presentation to the hospital, the responsible attending often chose to forego an ophthalmologic consultation. This occurred, for example, in many patients with fever, bronchiolitis, whooping cough, or other respiratory infections who were brought to the hospital after their caretakers observed a sudden episode of apnea, cyanosis, or limpness. None of the patients with shaken baby syndrome fell into this unexamined group, but it is possible that some of the unexamined patients may have harbored ophthalmologic abnormalities. Since we cannot assume that unexamined pa-

tients were entirely similar to examined patients, our data cannot be used to infer diagnosis-specific prevalences of ocular abnormalities in an unselected group of patients with ALTE. This caveat notwithstanding, the main conclusion of our study – that an ocular examination should be included in the evaluation of patients with unexplained ALTEs – would not change if ocular examination data had been available for all patients in the larger study.

The infant who has experienced an ALTE presents a challenging diagnostic problem. While patients with ALTEs often prove to have benign conditions, these events can also signal serious underlying pathology. One cause of an ALTE – shaken baby syndrome – can be especially difficult to recognize because the patient may appear well on presentation with no external signs of abuse. Because ophthalmologic findings can be critical in detecting this type of injury, physicians caring for patients with ALTE must consider obtaining ophthalmologic consultation early in the evaluation, and ophthalmologists must be aware of their potentially life-saving role.

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Reprint requests to:
Robin L. Altman, MD
Department of Pediatrics
New York Medical College
Valhalla, NJ 10595, USA
robin_altman@nymc.edu

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