

The First Cause of Traumatic Death in Children < 1 Year: A Review on Abusive Head Trauma

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Abstract

Context: Pediatric abusive head trauma (AHT) can be defined as an injury to the skull or intracranial contents of a child under the age of 5 due to inflicted blunt impact or violent shaking.

Evidence Acquisition: AHT is the most common cause of traumatic death in children younger than 1 year, and it is the leading cause of death due to child abuse. Clinical presentation observed in children with AHT depends on the type of AHT and accompanying injuries. History and physical examination are important for diagnosing AHT and for distinguishing it from other conditions that can mimic shaken baby syndrome, such as accidental trauma, cancer, metabolic diseases, and others.

Results: Progress in research on the medical diagnosis of AHT has been remarkable, while the development of treatment strategies has been limited. For these reasons, there is an urgent need to develop effective treatment strategies for AHT able to improve the outcomes.

Conclusions: The construction of a nationwide database that supports clinical studies is required in the future.

Keywords: Head Trauma, Shaken Baby Syndrome, Children

1. Context

Pediatric abusive head trauma (AHT) can be defined as an injury to the skull or intracranial contents of a child under the age of 5 due to inflicted blunt impact or violent shaking. Unintentional injury resulting from neglectful supervision, gunshot wounds, stab wounds, or penetrating trauma are excluded from the cause definitions. Shaken baby syndrome (SBS) is a type of abusive or non-accidental head injury caused by shaking alone or combined with an impact. It is more frequent in babies under the age of 1 year (1). The aim of this study is to provide a comprehensive overview of AHT in order to assist pediatricians in their clinical practice. Moreover, early diagnosis and accurate reporting and registering of AHT can reduce the morbidity and mortality associated with this condition, while missing the diagnosis can have severe consequences for the patient, also increasing the likelihood of recurrence (1, 2).

1.1. Epidemiology

AHT is the most common cause of traumatic death in children younger than 1 year, and it is the leading cause

of death due to child abuse (3). For several reasons, it is difficult to determine the actual incidence of AHT, and it is likely more common than hospital data suggest. First, not all abused infants need medical help and come into contact with the medical system. Second, not all cases presented in hospitals are diagnosed as cases of AHT. Third, the incidence varies depending on the data source (parental reports, national hospital discharge data, or fatality data) (4). Several prospective studies have been performed to try to establish the incidence of AHT. Most of these studies found that AHT is a relatively common cause of childhood neurotrauma, with an estimated incidence of 14 - 40 per 100,000 children under the age of 1 year: it is probably as prevalent in young children as are neonatal meningitis (25 - 32 per 100,000 live births) and lymphatic leukemia (28.7 - 36.6 per 100,000 children < 1 year) (5-7). Furthermore, most short-term outcome studies report high mortality and morbidity rates, with an additional cost of hundreds or thousands of dollars of excess inpatient, outpatient, and drug costs per patient for multiple years after AHT diagnosis (8).

1.2. Etiology and Risk Factors

The baby's crying pattern has been reported in the literature as the main trigger for the occurrence of AHT because a caregiver or perpetrator tries to stop the infant from crying by violent shaking. Crying is an important way for babies to communicate their needs (especially for newborns that are entirely dependent on their caregivers) and plays an important role in ensuring the survival, health, and development of the child (9). Brazelton has carried out an important study of baby's crying pattern: during the first weeks of a baby's life, there is an increase in the crying's duration, reaching a peak at 6 weeks, followed by a progressive decrease after the fourth month of life (10). The incidence of AHT strictly relates to the incidence of early infant crying. Inconsolable crying triggers a growing cascade of frustration and anger that results in decreased confidence in parenting ability (11).

Other situations known to trigger shaking are toileting and feeding difficulties (12).

Further known risk factors for AHT/SBS are:

- Male gender of the baby,
- Prematurity of the baby,
- Children from multiple pregnancies,
- Age under 1 year,
- Young maternal age (≤ 21 years),
- Male gender of the caregiver,
- History of alcoholism, drug abuse, or family violence,
- Parents' social isolation,
- Children residing in households with unrelated adults (2, 7, 13, 14).

Although the majority of known perpetrators are the child's father or the mother's partner, babysitters or carers are involved in about one case in five of shaking. Shaking occurs in all socioeconomic, cultural, and intellectual settings (2). Moreover, race and ethnicity have not been associated with a higher risk of AHT. Children in households with a single parent and no other adults in residence had no increased risk of inflicted-injury death (14).

2. Evidence Acquisition

AHT is characterized by several accelerations and decelerations with or without an impact. Injuries occur from the rapid and repetitive flexion, extension, and rotation of the head and neck around the torso. The perpetrator typically holds the infant by the chest, arms, shoulders, or legs and rapidly shakes the victim for 5 to 20 seconds. After the shaking episode, victims may be thrown against a surface (15). Babies are particularly susceptible to AHT because of

their large and relatively heavy head for their body and because their neck muscles are weak and undeveloped, allowing for substantial rotational shearing forces to be generated by shaking (9, 12, 16, 17). Moreover, an infant's brain has higher water content and less myelination than an adult brain does: indeed, it is easily distorted and compressed within the skull during a shaking episode. The lag time between the movement of the skull and skull contents, as well as any direct trauma, stresses and tears the blood vessels. The injured vessels start bleeding and lead to subdural hemorrhages, the most common intracranial finding associated with SBS (15).

2.1. Clinical Findings

Clinical presentation observed in children with AHT depends on the type of AHT and accompanying injuries. Brain swelling, subdural hemorrhage (SDH) and retinal hemorrhage (RH) are three classic symptoms that can indicate that an infant has been abused (18). Infants who have been shaken may present with or without neurological manifestations, including altered state of consciousness (77%), seizures (43% - 50%), vomiting (15%), and delayed development (12%). Although seizures are more common in AHT, this trend did not achieve statistical significance (19).

2.1.1. Brain Swelling

Brainstorming damage is multifactorial and occurs as a result of biomechanical forces; swelling, ischemia, and altered vascular auto-regulation contribute to the injury. In addition, axonal damage frequently occurs in deeper regions of the brain. The extent and severity of injuries depend upon the force and severity of the shake, the time span of the shaking, and whether an impact has occurred. Increased intracranial pressure is secondary to SDH, cerebral edema, and increased blood flow from cerebral injury (15).

2.1.2. Subdural Hemorrhages

SDH, described in 77-89% of patients, is the most common neuroradiological finding, even though it is not pathognomonic of shaking (20). With any rapid to-and-fro motion, the brain and bridging superficial cortical veins move at a different rate to the calvarium and attached dural venous sinuses. As a result, the rupture of cortical veins may create a hematoma in the subdural space (21). SDH suggestive of AHT is multifocal, located at the falx cerebri or posterior fossa and frequently accompanied by subarachnoid hemorrhage (2). Some authors reported that homogeneous hyperdense SDH at CT scan is significantly more common in cases of accidental head trauma

and that heterogeneous or mixed-density SDH is significantly more common in cases of AHT. However, mixed-density hematoma is not specific for AHT, because it may be observed within 48 hours of accidental injury (22). Children can present with a low level of consciousness, increased cranial pressure, seizures, apnea, hypotonia, anemia, and/or shock (23). SDH, frequently seen in SBS victims, can also result from accidental trauma, birth trauma, metabolic diseases, tumorous conditions, genetic disorders, and autoimmune disorders (24).

2.1.3. Retinal Hemorrhage

RH, rarely described in non-abusive head trauma (nAHT), is a cardinal manifestation of AHT: it occurs in 85% of cases of AHT, and its severity correlates with the severity of brain injury (25). The forces of a shaking episode may cause the layers of the retina to slide across each other, creating stretching and shearing of the retinal vessels, with hemorrhages (26). The vitreoretinal traction generated by the shaking seems to be the main cause of the hemorrhages, whereas the raising of the intracranial pressure plays a marginal role in the genesis of the lesions (2). Hemorrhages of the ora serrate are a specific sign of SBS: circular hemorrhages in the deep layers of the retina are called dot and blot hemorrhages. Blot hemorrhages are bigger than dot hemorrhages (15). The number, type, and distribution patterns of RH have significant diagnostic value. Typically, RHs associated with AHT are multiple, bilateral, confluent, multilayered (involving both preretinal and intraretinal layers), and extended from the posterior pole to the far retinal periphery (27). Moreover, the presence of retinoschisis and retinal folds in an infant with brain injury may be diagnostic of shaking (25). In contrast, RHs described in children with accidental head trauma are rare, more often few in number, and unilateral, involving only the retinal layer and located at the posterior pole (25, 28). An ophthalmologist should be consulted within 24 hours if an AHT is suspected. The indirect ophthalmoscope, preferably through dilated pupils, provides a three-dimensional view of the retina, allowing for an adequate examination of the posterior pole (27, 29). Differential diagnosis of RH also includes systemic conditions such as birth trauma, anemia, coagulopathy, increased intracranial pressure, leukemia, meningitis, hypernatremia/hyponatremia, and thrombocytopenia. A great number of ocular entities may be also associated with AHT, such as retinopathy of prematurity, hemangioma, and cytomegalovirus retinitis (29). Other ocular evidence of shaking injury may include optic nerve sheath and intraocular hemorrhages, as well as hemorrhages in the orbital fat or in extraocular muscles (25, 30).

2.1.4. Other Neurological Damages

Neurological manifestations also include injuries to the spine: for a long time, diagnosis of SBS was conducted by looking mainly for SDH, RH, and fractures; the progress in imaging techniques and the systematic dissection of the spine at autopsy after an infant death have shown the prevalence of these spinal injuries affecting not only the cervical spine but also the lumbar region (31). Epidural cervical hemorrhage and focal axonal damage to the brain stem and spinal nerve roots are the consequence of cervical hyperextension/flexion (32). Cervical trauma may lead to cardiorespiratory arrest and explains the sometimes-seen thin-layered subdural hematomas. Other neuroradiological findings include subarachnoid hemorrhages (12% - 25%), intracerebral hemorrhages (8%), epidural hemorrhages (4%), parenchyma lesions (37%), and hygroma (11%) (19).

2.1.5. Bone injury

The squeezing of the thorax is frequent, and the consequent rib fractures are powerful indicators of abuse. Most of them are localized in the back, near the vertebral articulation. Fractures of the costochondral junction are mainly localized in the lower chest. If a fracture of the “popping cribs” is found, then the liver lesions are often associated. Rib and chest lesions are more frequently observed in children at the age of 4 years, while children of less than 1 year of age are more often victims of cervical spine and ligamentous damages (15). Bone injuries suggestive of abuse also include signs of costal callus formation, metaphyseal fractures, humerus fractures, and periosteal spurs (2).

2.2. Diagnosis

History and physical examination are important for diagnosing AHT and for distinguishing it from other conditions that can mimic SBS, such as accidental trauma, cancer, metabolic diseases, and others (33).

2.2.1. Medical History

It is important to collect a comprehensive history of child abuse, but it is often unreliable and difficult to obtain. Parents and caregivers frequently withhold or misrepresent the history of the injury, so the abuse will be undetected. Historical indicators of physical abuse include:

- Delay in seeking medical assistance,
- Lack of an explanation for the symptoms,
- History inconsistent with the injury,
- Prior consultation for crying,
- History of unexplained sibling death,
- “Doctor shopping”,
- Conflicting histories given by caregivers (2, 34).

2.2.2. Physical Examination

A careful physical examination should be multidisciplinary and include research of signs and symptoms of abuse. Despite an accurate clinical examination, AHT still remains a difficult diagnosis to confirm in less severe cases (9,35). Strong evidence highlights the importance of investigating bone fractures, neck muscle damages, torn labial frenum, and abdominal trauma (36).

2.2.3. Laboratory Tests

A complete blood count and coagulation tests should be performed in order to complete the differential diagnosis with nAHT (34). A metabolic evaluation of the bone is also important: calcium and phosphorus metabolism, vitamin D levels, and the alkaline phosphatase levels should be dosed in order to exclude an osteogenesis imperfecta. In addition, a urine exam is relevant to find calciuria and phosphaturia, associated with other bone pathologies. Creatine phosphokinase should be checked to identify muscle damages such as contusions. Transaminases are elevated if a rib fracture has been caused by the shaking of the victim (37).

2.2.4. Imaging Studies

As there is no gold standard diagnostic test for child abuse, the diagnosis relies on clinical and radiographic features (38). A traditional X-ray examination of the head and neck can find multiple fractures; however, none of them is pathognomonic of abuse. Most fractures, abusive or not, are linear. Bilateral, multiple, or depressed fractures or fractures with a diastasis of more than 3 mm are more suggestive of AHT. Head X-ray may also help to rule out systemic illnesses such as wormian bones, osteogenesis imperfecta, or Menke's illness. An X-ray of the whole skeleton is useful in suspected AHT to search for other fractures that may be a sign of an underlying pathology (39, 40). The American Academy of Pediatrics guidelines recommends X-rays of the whole skeleton in association with bone scintigraphy to show evidence of bone lesions that are not visible on X-ray imaging (40). Brain ultrasound is indicated for children under 1 year of age when the cranial circumference is raised and an enlargement of the subarachnoid spaces is suspected. SDH can also be found. The CT scan of the brain remains the gold standard of traumatic lesions: it is time efficient, precise, and easy to use. In the acute phase, it is important to evaluate the necessity of a neurological intervention. A CT scan without contrast is sensible for subdural hemorrhages but cannot distinguish their abusive or accidental nature. Epidural hemorrhages are a suggestive CT scan sign of accidental impact trauma. In cases of doubt and if the result of the CT scan is initially normal, it could be repeated 12 or 24 hours later. MRI is of

significant diagnostic value and gives a complete overview of axial and extra-axial lesions. This should be performed as soon as the child's state has stabilized (2). The physical examination should also include an ophthalmoscopic evaluation (9).

3. Results

Most diagnoses can be differentiated from AHT by history, examination, laboratory, and/or radiologic evaluations. Accidents, birth trauma, and coagulopathies are among the most important entities to be excluded. The main differential diagnosis is accidental head injury. Short household falls from beds, sofas, and arms are common among children but rarely cause severe or fatal brain injury. Based on parental self-report, less than 1% of 3,357 fall in 2,500 infants aged 6 months or younger resulted in serious injury (41). Moreover, in a series of 287 children between the ages of 1 week and 6.5 years, SDH was found in 46% of children with AHT but in just 10% of children with accidental head injury (25). With regard to birth trauma, as many as 46% of healthy, term newborns have asymptomatic SDH, and RH may be present in up to 52% of infants, especially infants born of instrumented vaginal deliveries (42). All of these SDHs were resolved by between 4 weeks and 3 months later. These hematomas were differently located to SDHs with clinical manifestations. Typically, they present as a thin film of blood occipital or infratentorial overlying the cerebellar hemispheres (43). Congenital or acquired disorders of hemostasis, such as vitamin K deficiency, hemophilia, and thrombopenia, can present with intracranial, including subdural, hemorrhage during infancy (35). It is uncommon for bleeding disorders to present with intracranial hemorrhages, except for vitamin K deficiency (44). Although certain genetic and metabolic disorders can cause SDHs, most of the affected children will show other features as well. Genetic disorders include osteogenesis imperfecta, sickle cell anemia, Alagille syndrome, and Ehlers-Danlos syndrome. Although osteogenesis imperfecta is sometimes cited as a differential diagnosis, the overall clinical picture is very different, and a causal relationship with the presence of subdural hematoma has never been established. Metabolic disorders include Glutaricaciduria type 1 and Pyruvate carboxylase deficiency. Finally, in children with SDH without signs of trauma, infectious disorders (meningitis, Kawasaki disease, herpes simplex encephalitis, congenital toxoplasmosis) and intoxication (lead poisoning, cocaine, anticoagulant therapy) should be ruled out (5). Most of these diagnoses can be ruled out easily because they should have been known from medical history, are accompanied by other signs and symptoms (genetic disorders), or can be

rejected after simple laboratory investigation (coagulation disorders, infectious disorders). More difficult are cases where intracranial pathology is the only abnormal finding. AHT is the most common cause of SDH in young children, but in the absence of any other signs of trauma, establishing the diagnosis is more difficult for clinicians.

3.1. Management

Guidelines highlight that hospitalization is always indicated when the clinician suspects SBS (2). The identification and management of AHT require a multidisciplinary approach and coordination among health, child welfare, police, social services, justice, and education, as well as the community at large (12). Clinicians and pediatricians have the responsibility to report AHT to the competent authorities in order to protect the abused child and to prevent important neuropsychiatric sequelae. Unfortunately, few pediatricians diagnose AHT because of the social and legal consequences of that diagnosis: children could be removed from their homes, and parents could lose their parental rights or be imprisoned (40). A prolonged follow-up into adolescence and early adulthood should be systematically performed because the consequences of damage to the frontal lobe (responsible for behavioral problems) may not manifest until puberty or even later. Moreover, there is a high risk of potential long-term severe outcome in children initially thought to be normal on early follow-up (45). It is important to monitor development, detect any problem, and implement timely adequate rehabilitation interventions and/or support when necessary (46).

3.2. Prognosis

It has been calculated that approximately 1 in 250,000 children < 1 year will die from a short-distance fall. Another study found that the population-based risk of dying after a short distance fall for young children is less than 1 per million per young children per year (47). Many studies have demonstrated a statistically significant worse outcome (for both physical and cognitive functioning) for abusive head trauma patients than for accidental trauma patients (13). Niederkrotenthaler et al. (48) found that children with AHT were 5 times more likely to die and 8 times more likely to have a long duration of hospital stay than were children with nAHT. AHT has a poor prognosis: 15% - 23% of all recognized cases of AHT die before or shortly after presentation (7). Barlow et al. (45) documented that the morbidity rate in survivors was 68. A wide range of neurological sequelae (motor deficits, visual deficits, epilepsy, speech and language abnormalities), behavioral abnormalities (self-injurious and self-stimulatory behaviors, hyperactivity, impulsivity, temper tantrums, rage reactions), and sleep problems were observed.

4. Conclusions

As RH cannot always be detected on traditional ophthalmoscopy, a technique with more sensitivity and specificity is required. The Ret Cam is a digital color fundus camera with a contact fiber-optic, providing 120° field imaging that enables rapid capture of eye fundus images. A pilot study has compared Ret Cam imaging for telemedicine to standard ophthalmoscopy to study the RH. The presence or absence of RH was assessed by both methods, but the digital camera could detect the retinal abnormalities in all cases examined, against 67% for ophthalmoscopy. Therefore, remote reading of Ret Cam photographs could be a promising future strategy for detecting children with AHT (49). A psychosocial team should work with pediatricians to evaluate social risk factors of AHT and protection measures (13). Progress in research on the medical diagnosis of AHT has been remarkable, while the development of treatment strategies has been limited. For these reasons, there is an urgent need to develop effective treatment strategies for AHT able to improve the outcomes. At the same time, the construction of a nationwide database that supports clinical studies is required in the future.

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