The shaken baby syndrome

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ABSTRACT

Trauma is the most common cause of death in childhood and non-accidental injury is the leading cause of death in infants between one month and one year of life. This is a newly emerging entity in Saudi Arabia. However, there is little available literature on the extent of child maltreatment in Saudi Arabia and other Arab countries. In this review, we will discuss various aspects of the central nervous system insults resulting from the inflicted trauma of child abuse. We aim to raise awareness in the region as the tragic loss of life and function is unequalled in childhood beyond the perinatal period.

Keywords: Injury, shaken, baby, infant, non-accidental.


Trauma is the most frequent cause of death in childhood. Non-accidental injury is the leading cause of death in infants between age one month to one year in North America. With respect to the central nervous system, the inflicted trauma of child abuse (non-accidental trauma) produces high morbidity and mortality. Child abuse has long been recognized. In children less than one year of age, child abuse accounts for over one third of all craniofacial trauma and 95% of intracranial hemorrhages. A disproportionate 80% of deaths in children beneath two years of age results from non-accidental trauma despite the fact that non-accidental trauma occurs less frequently than accidental injuries. In Canada, physical assault suffered by children most often injures the child's head and brain; nearly one half of the victims of such violence are infants or children less than a year old. The clinical coincidence of subdural hematomas and characteristic metaphyseal avulsion fractures in traumatized children was first reported by Caffey in 1946. Subsequent to Kempe et al in 1962, description of the "battered child syndrome" a modern literature arose documenting the spectrum of abuse suffered by children at the hands of others.

Fatal non-accidental trauma (homicide) accounts for 17% of all deaths in children aged less than one year. Furthermore, homicidal injuries cause 5% of all childhood deaths in comparison to 1% of all adult deaths. A study of childhood homicide in Northern England reported 49% of homicide victims died following blunt injuries, and 34% of the victims had previously been abused. Most disturbing was the fact that 33% of these homicides occurred in children less than 1 year of age. In this group, intracranial injuries caused 60% of the deaths and 62% of the victims had suffered abusive trauma prior to lethal injury. In the face of such dismal statistics in children incapable of defending themselves or even avoiding injury, prevention is the only principal means capable of changing this tragic situation. Physical violence is never an acceptable punishment to a small child or infant.

Child abuse is a newly emerging entity in Saudi Arabia. However, there is little available literature on the extent of child maltreatment in Saudi Arabia and other Arab countries. Few case reports and series addressed this problem in Saudi Arabia. Al-Ayed et al, found the incidence steadily rising, however, the total number was very low. It is possible that this problem is underdiagnosed and the rising incidence reflected improved case recognition.
A recent study conducted in Egypt revealed that children are subjected to physical abuse in a disciplinary context. Moreover, schoolteachers, social workers, and physicians do encounter cases ranging from neglect to sexual abuse. A more recent community-based study from Alexandria investigated mother's behavior to child abuse and how it correlates to parent and family characteristics. Child abuse was significantly associated with parent's educational level, parent's occupation, financial problems, crowding index, and maternal perception of maltreatment.

In this review, we discuss various aspects of the central nervous system insults resulting from the inflicted trauma of child abuse. We aim to raise awareness in the region as the tragic loss of life and function is unequalled in childhood beyond the perinatal period.

**Non-accidental Central Nervous System (CNS) injury.** Variously termed the whiplash-shake syndrome, the shaken infant, the shaken-impact syndrome, or the shaken baby syndrome. Non-accidental central nervous system injury in infants (shaken baby syndrome) has been recently reviewed. It presents characteristic signs readily apparent only to those who suspect it may have occurred. These include: Altered level of consciousness in an infant or toddler, retinal hemorrhages, radiographic evidence of characteristic intracranial hemorrhages (e.g., intrahemispheric subdural hematoma) or brain swelling.

Reflection, careful history taking, and thorough examination typically confirm the above findings while documenting a vague uncertain, or absent history inconsistent with the injury or disability evident upon examination. Clinical presentation may suggest seizures, post-ictal depression, respiratory or cardiovascular collapse, or infection. External signs of trauma are commonly minimal or absent. Irritability and a depressed level of consciousness may be the only findings present upon initial examination.

Prominent external signs of trauma to the face, head torso or limbs, imply direct injury by blunt trauma or other means, but do not eliminate the possibility of coincident shaking or shaken-impact injury. Minimal or absent signs of external trauma are typical of shaken-impact injuries. A thorough search for retinal hemorrhages must be pursued early during examination once vital signs are stable and adequate airway and oxygen provision are assured. Provisional diagnosis and appropriate diagnostic investigations are determined by these initial clinical findings.

**Clinical assessment. History.** All too frequently, important details of history are absent or vague and confusing. Lacking a history consistent with the physical findings, an infant or toddler with an altered level of consciousness must be presumed at high risk for non-accidental trauma. To be appropriate the history must be consistent with the findings of subsequent examination and diagnostic studies. Infants and toddlers who suffer minor falls do not sustain major skull fractures or severe depression of consciousness in routine circumstances. Severe cranioencephal trauma in children of this age most often results from falls from significant heights, automobile accidents, or less frequently, falls down a flight of stairs while in a baby walker. Such severe accidental injuries are typically witnessed by others who provide specific consistent historical information. In contrast, when available, the history of non-accidental trauma is imprecise, may change from one interview to the next, or may be misleading or false. Nevertheless, the available history should be accurately documented.

**Physical examination.** Due to the available history that is often false or misleading, the initial examination is a critical element in the abused child's assessment. As the child's vital signs are assessed, a rapid search for external signs of trauma is completed. Vital signs are stabilized if necessary and respiratory adequacy assessed. Abnormal respiration or hypoventilation suggests severe CNS injury may be present. Hypotension should prompt administration of intravenous fluids and appropriate supportive measures. If emergency intubation is necessary, a rapid neurologic assessment, including pupillary evaluation and assessment of Glasgow Coma Score, should be completed during 100% oxygen ventilation before pharmacologic paralysis and intubation. The presence and character of the anterior fontanelle are assessed. A tense fontanelle suggests intracranial hypertension, but says nothing regarding the cause of such elevated ICP. Meningismus or opisthotonus also suggest CNS damage or intracranial hypertension. A deliberate search for retinal hemorrhages must be carried out, early detection of significant retinal hemorrhages does not require dilution of pupils. Neurologic assessment may then be completed along with the remainder of the physical examination. Potential pitfalls in the assessment of infants include: Missed retinal hemorrhages, inappropriate Glasgow Coma Score parameters, inappropriate assessment of Glasgow Coma Score.

Retinal hemorrhages, classically the hallmark of the shaken baby, are noted in 50-100% of the reported shaken children and when present, are almost pathognomonic of this form of child abuse. Careful fundal examination is needed in any infant presenting with acute CNS dysfunction, including seizures to identify hemorrhages. Retinal hemorrhages should prompt computerized tomography (CT) imaging of the central nervous system as soon as any needed resuscitative measures are completed and vital signs are stabilized. When retinal hemorrhages are absent, a broader spectrum of diagnoses must be considered.
but the need for CT imaging is not necessarily lessened. If CT is not carried out, the extent or cause of CNS injury may not be readily appreciated and thus appropriate treatment may be delayed.

Professionals who do not frequently assess injured infants may misinterpret stereotypic activities as volitional or spontaneous events. Apparently, spontaneous repetitive eye opening may be seen in infants with significant cortical injury. Spontaneous bicycling movements of the legs or the arms may occur despite severe brain injury. As Bruce suggests, evaluation of the level of consciousness in the infant less than six months of age should rest upon eye opening, ocular tracking, head control, crying, sucking, observed motor activity, and developmental reflexes.\(^2\) Pupillary reflexes, oculocephalic reflexes, gag, and respiration assess the status of the brainstem and medulla.\(^3\) The comatose infant does not cry.\(^10\)

Lastly, hypotension in infants and toddlers suggests injury to the cervicomедullary junction and to an abdominal vescus or both. Cervicomедullary injury may yield apnea and flaccid paralysis in addition to hypotension and cardiac instability. Blunt abdominal trauma may produce significant peritoneal or retroperitoneal hemorrhage, or peritonitis.

**Diagnostic studies.** Initial diagnostic studies should include routine blood samples, electrolytes, oxygen saturation measures (e.g. venous, capillary or arterial blood gases) and appropriate radiographs (Skeletal survey, bone scan to identify fresh fractures). For legal reasons the following tests are also needed to rule out blood disorders (PT, PTT, CBC (plate), bleeding time). Subsequent imaging studies (e.g. CT) are determined by physical examination findings and the examining physician's assessment of the overall clinical situation.

**Missed diagnosis.** Many inflicted injuries are misinterpreted as accidents or mistaken for other neurologic disorders.\(^1\) If misled by false or vague history, or should the lack of history or key physical findings confound an examiner.\(^25\) The following sentinel signs guard against missed diagnoses in the case of shaken-impact abuse: Retinal hemorrhages reliably indicate a traumatic etiology that should be presumed non-accidental until proven otherwise. Cardiopulmonary resuscitation and accidental trauma virtually never cause significant retinal hemorrhages.\(^21,22,24,26-28\)

Bloody cerebrospinal fluid obtained upon lumbar puncture suggests intracranial hemorrhage (subarachnoid hemorrhage or subdural hemorrhage). Simple interpretation as “traumatic tap” risks a missed diagnosis and may result in a fatal outcome.\(^29,30\) Computerized tomography demonstration of parafalcine occipitoparietal intrahemispheric subdural haematoma is indicative of shaken-impact injury.\(^2,20,31,32\)

**Clinical imaging studies. CT Imaging.** Cranial CT is the best initial imaging study for the evaluation of childhood non-accidental CNS trauma. Plain skull radiographs may be obtained, but in the situation of significant head injury they add little to solve the diagnostic puzzle. The severe CNS injury may occur coincident to, or in the absence of a skull fracture. Furthermore, where clinical assessment or caution suggest, the CT examination may easily be extended to include the spine, abdomen, pelvis, or chest.

The most common CT lesion documented in shaken-impact injury to child abuse is a parafalcine subdural hematoma located in the occipitoparietal region, often with extension into the interhemispheric fissure. The attendant mass effect may relate to the subdural blood or to the underlying parenchymal injury. Some consider this injury characteristic for this mechanism of child abuse.\(^2,20,31,32\) Intracranial hemorrhage may also occur at other sites.

Subdural or intracerebral bleeding is often accompanied by traumatic subarachnoid hemorrhage (SAH). Cerebral swelling, evidenced by regional or generalized hyperlucency with poor definition of the gray-white matter interface normally seen in each hemisphere, is another frequent CT finding. This is often coincident with the posterior subdural hematoma described above. The loss of gray-white matter distinction and hypolucency are the result of ischemia or infarction. Infarction or ischemia may result from tearing veins bridging the gap between brain and sagittal sinus, from the same injury that produced the subdural hematoma or other laminar subdural fluid collections, or may be the result of other factors. When present bilaterally, it suggests hypoxic-ischemic injury, which may be secondary and related to apnea or hypotension produced by the primary traumatic injury. Cerebral swelling may be attended by significant mass effect with midline shift, displacement or compression of ventricular structures, or in the case of herniation, displacement or obliteration of basal cisterns.

Computerized tomography may also visualize other injuries such as fractures (especially depressed) or indirect signs of increased intracranial pressure (e.g. split cranial sutures). Computerized tomography imaging often readily demonstrates significant pathology, however, it does not always accurately reflect intracranial hypertension. Follow-up CTs may demonstrate progressive changes in subdural collections or brain parenchyma. Repeated scans are appropriate when the clinical condition changes significantly or when appropriate treatment fails to yield an expected result.

**Magnetic Resonance Imaging.** Though inappropriate in the acute setting, magnetic resonance imaging (MRI) may assist in the diagnosis of difficult child abuse cases,\(^25\) or in the assessment of CT documented injuries. Magnetic resonance imaging can detect small subdural hematomas, cortical contusions, or deep white matter lesions, and display them with exquisite anatomic detail. Such
detailed resolution of the true extent of injury may allow more accurate prediction of outcome. Magnetic resonance imaging also clearly demonstrates progressive dissolution of hemoglobin and methemoglobin allowing an estimation of the time of injury and the detection of multiple episodes of trauma (e.g., laminar densities in a "mixed" subdural hematoma, or hemorrhages of different ages). Though not yet cited in the literature, the sensitivity and resolution of magnetic resonance imaging may allow the detection of other subtle injuries not currently revealed by high resolution CT. The detection of injuries to the galea, medulla, or cervical spinal cord could be critical factors allowing proper clinical and forensic correlation of potential mechanisms of injury and improved prediction of outcome after non-accidental CNS trauma.

Pathophysiology of non-accidental CNS injury. A detailed discussion of the spectrum of mechanisms and associated pathophysiology of non-accidental CNS injury exceeds the scope of this paper. Metabolic pathologic investigations of both clinical and experimental trauma suggest at least three potential mechanisms may be pertinent: Impact trauma related to direct injury (blunt trauma). Impulsive injuries related to shaking alone (whiplash-shaking syndrome). Impulsive injuries initiated as shaking and terminated with impact (shaken-impact syndrome). Which of the latter two mechanisms predominates remains a controversial topic, but each of the three syndromes may occur in relatively "pure" form.

Direct or blunt trauma is typically associated with obvious external trauma to the face, scalp, and cranium producing fractures and direct brain injury in addition to axonal shear injuries. When external signs of trauma are minimal or absent, shaken-impact abuse, as proposed by Duhaime et al. is likely the most common mechanism of injury. Less commonly, shaking alone (the whiplash shaking syndrome) described by Caffey is the causative mechanism. Injuries to important brainstem centers (e.g., locus ceruleus), medulla, and cervical cord commonly occur in each shaking mechanism. This rapidly produces apnea or impaired respiration, cardiac instability, hypotension, and loss of consciousness. Lesions characteristic of diffuse axonal injury are prominent features common to all three mechanisms. Infants and toddlers are uniquely susceptible to injury by shaking abuse. The child’s large head and minimally myelinated brain are poorly protected by a thin pliable skull and poorly supported by a hypermobile spine with weak stabilizing musculature. A relatively high brain water content, an average cerebral blood flow twice that of adults, and generous subarachnoid spaces are additional factors that may potentially aggravate accidental or abusive trauma.

Critical lesion sites. A brief listing of lesions occurring at sites critical to the proper understanding and diagnosis of abusive CNS trauma in young children must include: Scalp or galeal contusions, indicative of a direct impact component of injury. Gliding contusions (parasagittal cortical lesions), indicative of acceleration-deceleration injury. Intracranial hemorrhages, characteristic lesions designated previously (e.g., subdural hematomas) may also be accompanied by isolated or multiple cortical contusions. Diffuse axonal injury (e.g., scattered parenchymal petechial contusions), clinically presents as a major alteration of level of consciousness and focal neurologic deficits or both. Corpus callosum contusion or tears (microscopic or macroscopic), with the latter perhaps visible on imaging studies. Contusions or hemorrhage within the dorsolateral pons and brainstem or both may disrupt important centers mediating consciousness and cardiovascular homeostasis. Secondary Duret hemorrhages are not seen in infants and toddlers. Therefore, brainstem hemorrhages are the direct or primary result of trauma, occurring prior to the onset of cerebral swelling and intracranial hypertension. Cervicomedullary junction contusion, tear, or hemorrhage may result from whiplash-shaking alone or from shaking impact injury, and may produce in rapid respiratory death and flaccid quadraparetic coma or both. Cerebral swelling, ischemia, or infarction may be regional or general, affecting one or both hemispheres. The significance and characteristic features of these critical lesions are appropriately reviewed and discussed in references.

Ocular lesions. Ocular hemorrhages and trauma coincident to child abuse are, like the proposed mechanisms of abuse, a mix of accepted principle and published controversy. In a population limited to infants and toddlers, retinal hemorrhages are readily accepted as diagnostic hallmarks of abusive trauma. Other ocular pathology, such as retinal or macular folds, which may in the future prove to be specific sequelae of abusive trauma have been reported. The exact mechanism by which retinal hemorrhages arise, whether they may represent a secondary injury, and the frequency with which they may be seen in nonabusive or accidental trauma remain prominent points of controversy. Reported late ophthalmic consequences of abuse by shaking include impaired visual acuity, visual deficits related to retinal or macular folds, optic nerve lesions, and cortical blindness. Visual inattention or neglect may complicate recovery from injuries to association cortex or sites not involving traditional visual pathways.

Principles of treatment. Non-accidental CNS trauma seldom produces lesions requiring surgical treatment (e.g., depressed fractures, large subdural hematomas, or rarely significant epidural hematomas). Surgical lesions generate secondary damage superimposed upon the initial traumatic

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injury, and are typically related to direct impact, frequently occurring within the context of multiple injuries to the central nervous system, abdomen, chest or extremities.

In contrast to direct impact injury, abuse by whiplash-shaking or shaken-impact mechanisms characteristically produces primary CNS damage that persists despite treatment. Secondary factors such as reactive cerebral swelling, ischemia, hypoxia, seizures, or disrupted electrolyte maintenance require aggressive medical management in an intensive care setting. Progressive intracranial hypertension invariably accompanies severe non-accidental CNS damage. Patients who remain comatose or who neurologically deteriorate require ICP monitoring once CT demonstrates no surgically amenable lesion is present. Adequate control of increased ICP by means of controlled ventilation, elevation of the head, relative hypocarbia, optimal systemic and cerebral perfusion pressures, diuretics, sedation, and where necessary, the use of steroids or barbiturates, is the goal of treatment. Detection and treatment of seizures is important as well, especially in the sedated and paralyzed or comatose patient who may suffer unrecognized or subclinical seizures detectable only by electroencephalographic monitoring. While the primary damage produced by shaking or abuse cannot be reversed, any appropriate treatment that may prevent or remedy additional injury by secondary factors should be pursued.

Most deaths in shaken infants are the result of intractable intracranial hypertension a summation of the primary and secondary insults occurring before diagnosis and the onset of appropriate treatment. Those children who succumb typically present in deep coma with apnea or impaired respiration, demonstrate severe abnormalities on motor examination, have poor delineation of gray/white matter on initial CT imaging, and have a very high ICP at the time treatment commences.²

**Outcome.** Infants and toddlers who present with obvious alteration of consciousness, minimal or no external signs of trauma, retinal hemorrhages and CT findings typical of shaking abuse have a poor prognosis. One third die from their abuse, one third remain significantly disabled by mental, cognitive or physical impairments, and the remaining one third have a “good” recovery but do not necessarily become normal in all respects. Overall, these prospects are worse than those following severe vehicular trauma. Most of the children who die from their injuries are near death before their true condition is recognized or medical assistance is sought. Survivors of abuse remain at high risk for repeated abuse or death unless their plight is recognized and they are removed to a safe non-abusive sanctuary.²⁶⁴³ Prevention of abuse by education of families and caretakers, and if necessary by physical removal of the victim to an abuse-free environment remains the only appropriate resolution for violence directed against children. Treatment is woefully inadequate and hopes of cure remain unfulfilled.

### References


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