

Shaken Baby Syndrome as a Form of Abusive Head Trauma

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متلازمة الطفل المهزوز كنوع من أنواع إصابات الرأس الناجمة عن إساءة المعاملة

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المخلص: متلازمة الطفل المهزوز هي إحدى أشكال إساءة المعاملة التي تتميز بإصابات في الدماغ. تمثل المتلازمة معضلة تشخيصية للأطباء المعالجين بسبب عدم وضوح المشاكل العصبية والسلوكية للمصاب وإن كانت منهكة. ما يجعل الوضع أسوأ هو السن المبكرة للطفل المتضرر والتي تجعل الطفل غير قادر على شرح ما حدث. سُجّلت حالات متلازمة الطفل المهزوز في مناطق عديدة من العالم، لكن وحسب معلوماتنا هناك ندرة في الأدبيات حول هذا الموضوع في الدول العربية والإسلامية. هذا المقال يحاول تسليط الضوء على المتلازمة من خلال مراجعة المعلومات الخاصة عن المسببات المرضية لها، فضلاً عن التشخيص وأسباب تأخر التشخيص. الهدف الرئيسي من هذا العرض هو زيادة الوعي بمتلازمة الطفل المهزوز بحيث يمكن وضع السياسات المستنيرة للوقاية والتدخل في المنطقة وخصوصاً في سلطنة عُمان.

مفتاح الكلمات: متلازمة الطفل المهزوز؛ إساءة معاملة الأطفال

ABSTRACT: Shaken baby syndrome (SBS) is a form of abuse that is characterised by brain injury. Because of the subtle and yet debilitating neurobehavioural impairment which ensues, SBS represents a diagnostic dilemma for attending clinicians. The situation is made worse by the young age of the affected child who may not be capable of explaining what happened. SBS has been reported in many parts of the world. To our knowledge, there is a dearth of literature on the topic from Arab/Islamic countries. This article attempts to shed light on the syndrome by reviewing information on the aetiology of SBS, as well as on its diagnosis and the reasons for delayed diagnosis. The central aim of this review is to increase awareness of SBS so that enlightened policies for prevention and intervention could be developed in the region and particularly in Oman.

Keywords: Shaken baby syndrome; Child abuse

CHILD ABUSE IS A GLOBAL PROBLEM THAT presents in the forms of physical abuse, sexual abuse, neglect, and emotional abuse.¹ Children subjected to various form of abuses can develop emotional, cognitive and physical conditions. The magnitude of abuse in children is often underestimated either because the sequelae of the abuse, or their young age, makes the victims incapable of articulating what happened to them.²

Shaken baby syndrome (SBS) is one form of abuse that predominantly impacts neurological functioning, but it can also have multiple impacts on neurocognitive functioning. SBS is a relatively new diagnosis that was first described by Guthkelch in 1971.³ The first name used to describe it was

'whiplash-shaken infant syndrome'. Other terms used were 'shaken infant syndrome', 'shaken impact syndrome', 'infant whiplash-shake syndrome', 'abusive head trauma', and 'inflicted/non-accidental' or 'intentional head injury'. In medical literature, SBS is the most widely used and recognised term;⁴ however, the term is not widely accepted. For example, the American Academy of Pediatrics prefer 'abusive head trauma' which is broad and inclusive of all mechanisms of injury.⁵

The number of articles on SBS has significantly increased in recent years.¹ It is evident from the literature review that no publications on SBS have emerged from Arabian Gulf states. This region has a population structure with a large proportion of

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children and adolescents; however, despite rapid economic growth in the region, no child protection agencies have been established. Anecdotal and impressionistic observations reveal that something akin to SBS is seen in the region. The aim of this article is to synthesise some of the relevant literature and shed light on the intricacies of SBS.

RISK FACTORS AND CAUSES

The victims of SBS are usually under one year-old and thus helpless and unable to protect themselves.⁶ However, there are a few case reports of older children with closed head injury with severe neurologic impairment and ophthalmic findings suggestive of a shaking injury.⁷ Infants and children are particularly vulnerable to violent shaking because of their relatively large head and weak cervical musculature.^{8,9} These factors, together with the incompletely fused sutures and relatively large volumes of cerebrospinal fluid in young children, allow for greater movement within the cranial vault, resulting in potentially severe damage to the immature incompletely myelinated brain.^{8,9} Perceived disruptive behavior such as unwarranted and unremitting crying spells have been suggested as being important precursors of abuse in the form of 'shaking' by significant figures in the child's life.¹⁰

Perpetrators of SBS are often male with the biological father being the most common abuser.¹ The literature also suggests that stepfathers or male partners may also be involved in such heinous acts. Female babysitters are also known to be perpetrators of SBS, as well as biological mothers. Overall, the perpetrator was male in 72% of cases.^{1,11} There is speculation that males, due to their greater physical power, are more likely to cause SBS when they 'shake' children.¹ One might argue that females might be more tolerant to babies' needs and demands. In contrast, males are perhaps more easily provoked by a crying baby. On the whole, there is evidence to suggest that both parties contribute to this abuse. The distressed child may cause stress to a significant figure thus provoking them into abusing the child. There is also evidence to suggest that individuals who are prone to anger and marked by explosive personality disorder are likely to commit such acts.^{10,12}

MECHANISM OF INJURY

SBS is an extremely serious form of abusive head

trauma that occurs when a child is held by the torso or the extremities and subjected to violent shaking that results in rapid head movements with acceleration, deceleration and rotational forces, with or without impact. It results in a unique constellation of intracranial, intraocular and skeletal injuries.^{7,13} The most common intracranial abnormality detected is subdural haematoma. Children with SBS are often found to have retinal haemorrhage.^{2,11,14} Fractures of the ribs where the child is grasped, or long bone fractures when child is held, might also be detected. Cervical spine injuries are rarely recorded in cases of SBS.^{11,13}

Diagnosis

SBS is known to be difficult to detect and diagnose. Clinicians should use their own clinical judgment as each individual case is different and needs to be considered carefully on its own evidence. According to estimations from the World Health Organization (WHO), in 2002 almost 31,000 children aged <15 years died worldwide as a result of homicide.¹⁵ Despite advances in investigative neurology, abusive head trauma is commonly under-recognised and remains a diagnostic challenge. The diagnosis of SBS must be considered in any infant or young child who collapses with no obvious causes. Clinicians must maintain a low threshold of suspicion for considering this diagnosis.¹⁶ The diagnosis of SBS is usually made following a careful medical and social history taking. This ought to be supplemented by appropriate investigations. Children with SBS are often seen first at emergency departments (EDs). The incidence rates of child abuse at EDs ranges from 2–10%; the detection rate might increase if medical staff were systematically vigilant about the possibilities of abuse in each child they encountered.¹⁷

A comprehensive history of the presenting complaints is an essential component of the diagnostic process. Infants with SBS present to hospital with a variety of symptoms ranging from vomiting, poor feeding and lethargy to convulsions, apnoea and death. Symptoms occur immediately after the insult, thus recording the timing of the symptoms is very important.^{18,19} Not all infants are acutely ill at presentation, and in some cases the absence of either a history or external signs of injury may delay diagnosis. It is also important to note that

shaking alone without impact injury can produce the symptoms seen in children with shaken baby syndrome.⁹ There are a number of features in the children and parents that may increase suspicion that harm has been done; these are triggering factors such as “crying, temperamental behaviour, toileting problems” and a history of previous or recent injury.¹⁹ Birth history, developmental milestones, and vitamin K status are also important to note. In most cases where a history of injury is given, it is reported to be of a minor nature and is not consistent with the severity of the infant’s condition. A review of checklists of risk indicators for child abuse in emergency departments, shows that three history items are worthwhile considering: delay in seeking medical advice, an inconsistent history, and clinical findings that are incongruent with the history narrated by the accompanying adult.¹⁷

Physical examinations for SBS should include all of the body looking for signs of external injuries such as skin bruising, abdominal injuries and skeletal injury such as fractures of ribs or long bones.¹⁹ The child conscious level, neck and cervical spinal cord injuries should be evaluated together with the need for resuscitation. It is also necessary to look for signs of intracranial bleeding such as fullness of the fontanelle and increased head circumference. Fundi should be examined by the clinician/paediatrician, and as soon as possible by an experienced ophthalmologist, in order to exclude eye injury including retinal haemorrhage. Neuro-imaging is the definitive diagnostic investigation and should be performed whenever SBS is suspected. The first line investigation in suspected abusive head trauma is a computed tomography (CT) head scan followed by a magnetic resonance imaging (MRI) scan. MRI is a more sensitive method of detecting small intracranial collections, especially in areas less well seen on CT.⁴ Cerebral oedema and ischaemic changes are also well demonstrated by diffusion weighted MRI. A skeletal survey, including skull films, should be performed in all children less than three years old where physical abuse is suspected.¹⁹

The clinical diagnosis is usually based on a patient history that does not explain the clinical features; it is supported by the findings of the physical and retinal examinations and the brain MRI.

OPHTHALMIC MANIFESTATIONS OF SBS

Ocular manifestations of SBS, characteristically retinal haemorrhages, are seen in 85% of cases and are bilateral, diffuse and multilayered.²⁰ A large study from the USA concluded that retinal haemorrhages in young children are associated with a high likelihood of abuse.²¹ Other ocular manifestations of SBS include blood-filled schisis cavities and circumlinear perimacular folds.²² Papilloedema may also be noted. The anterior segments are usually unremarkable in SBS. The pathogenesis of the ocular findings is the same as the intracranial manifestations, namely repetitive, to and fro acceleration-deceleration forces, that cause a displacement of vitreous volume and a resultant traction on the retina and retinal vessels resulting in rupture and haemorrhage.

The presence of retinal haemorrhages implies a shaking aspect to the trauma; these are rarely seen in victims of road traffic accidents who have skull fractures and intracranial bleeding due to direct trauma.²³ Minor falls and blows, so common in the domestic context, do not cause retinal haemorrhages. Seizures are also not known to result in retinal haemorrhages.¹⁴ Differential diagnoses of retinal haemorrhages in this age group of children includes birth trauma, severe life threatening accidental head injury, coagulopathies, sepsis and vasculitis. With the exception of birth trauma, retinal haemorrhages in the other conditions tend to be few and localised.¹⁴

MANAGEMENT

Where abusive head trauma is a possibility, a strategy discussion involving police and the children’s social carer should be held to decide whether to initiate enquiries and then a criminal investigation. Children are frequently referred to a specialist centre where paediatric neuroscience resources are available. It is important that such specialists are supported by general paediatricians who are able to liaise with the local and statutory child protection teams and participate fully in procedures for safeguarding the child. Laboratory investigations are necessary in order to exclude other medical conditions such as rare metabolic diseases (glutaric aciduria), coagulation disorders, and infective encephalopathy.^{24,25} Other investigations should include a septic screen to exclude infection—as

subdural collections could be associated with meningitis, urine screening for toxicology, and a metabolic screen. It is also important to do a full blood count, repeated after 24–48 hours, which may demonstrate a rapidly falling and low haemoglobin level.¹⁶ The role of the ophthalmologist is often to assist in the diagnosis of SBS by examining the child for retinal haemorrhages. Retinal haemorrhages often resolve spontaneously and do not require therapy; however, extensive, non-resolving vitreous haemorrhage or retinal detachment might mandate surgical intervention. Careful follow-up is desirable to document and treat sequelae which may be consequent to neurologic or ocular damage.²⁶

OUTCOME

The available evidence suggests that around one third of severely shaken infants subsequently die as the result of being shaken.^{1,4,26} This rate is 6% to 12% higher than that for accidental head injuries in a similar age range.²⁶ The syndrome has an extremely high degree of morbidity with 60% of survivors having a moderate or greater degree of disability.¹¹ The victims of SBS display a wide range of neurological sequelae.²⁶ These include cognitive and behavioral disturbances, cerebral palsy, blindness, and epilepsy.⁴ It is well known that outcomes due to head injury are more severe in abused children compared to injuries resulting from unintentional accidents.^{2,27} The functional prognosis of children marked by SBS is likely to be poor. This partly reflects the silence on the part of the parent who may conceal the shaking incident since this might lead to legal prosecution. The victim themselves are likely to be rendered incapable of 'complaining'. The very fact that the incident is associated with neurological dysfunction, makes it likely that some residual though subtle impairment is likely to be chronic and have severe consequences in term of quality of life.^{28,29}

BIOMARKERS AS THE FUTURE HOPE FOR RAPID SCREENING OF NEUROTRAUMA

Many cases of brain injury can present with clinically non-specific symptoms such as vomiting, poor feeding and irritability.¹¹ It is worthwhile noting that such a presentation may be indistinguishable from other childhood maladies such as gastroenteritis or respiratory tract infection.¹¹ The history will be

misleading and clues to the cause of the head injury will be omitted. All of these factors can contribute to a delayed/missed diagnosis. Many research studies are underway to find biological makers of SBS. Despite the importance of such biological markers, there is, as yet, no study supporting their validity and reliability.³⁰ The clinician's vigilance still remains the best option.

DILEMMA OF SBS

The diagnostic dilemma of this syndrome is three-fold. The first is that the shaking is not witnessed and even when a perpetrator confesses the full truth of the incident most likely is not fully revealed. This results in questions about whether the shaking event is the cause of the clinical and pathological findings in the affected children.^{1,31,32} However, it is suggested that mechanical shaking is the primary reason for retinal bleeding as researchers found no difference in the extent, type, or frequency of retinal bleeding between patients diagnosed with SBS who exhibited signs of direct impact trauma versus those who did not.¹⁴ The finding of positive correlation between retinal haemorrhages and intracranial injury may support the theory that mechanical shaking and its direct effect on the globe and orbit has a major role in the pathogenesis of SBS.^{7,14} Such studies and other results support the view that shaking alone is able to produce the symptoms seen in inflicted traumatic brain injury.¹⁸

The second factor is the possible medical conditions that could have similar presentations to abusive head trauma such as birth and other accidental injuries, brain congenital malformations, genetic and metabolic conditions, hematological disorders, infectious diseases, toxins, complications of surgical intervention, and nutritional deficiencies.¹⁹ Short falls are the primary cause of injury given by care providers in most of the cases of SBS. Studies found that minor falls (< 4 feet) do not cause serious injury in children, except for epidural haematoma whereas subdural and/or subarachnoid haemorrhage are seldom seen and retinal haemorrhage are virtually never seen in short falls.² Such findings minimise the possibility of a short fall being the causative factor of the retinal and intracranial findings of SBS. The possibility of a neonate developing intracranial bleeding after normal delivery is not rare and the potential for serious injury following a fall from

a short distance is not common, but can happen. This necessitates consideration of these two possibilities when contemplating the diagnosis of abusive head trauma.^{11,33,34} Such medical mimics of SBS, compounded with an unreliable history, may make the diagnosis challenging, especially in children with absent external injuries as is the case in 40% of affected children.¹¹ The third factor is the variable results of biomechanical studies of SBS injury which have to take into account the levels of rotational velocity, acceleration of the head, velocity from manual shaking, the presence of hypoxia and the associated potential injuries to the brain and cervical spine.³⁵⁻³⁹

Conclusion

It is important to raise awareness about abusive head trauma among paediatricians and general practitioners in Oman. Doctors providing health services to children should be alert to the non-specific symptoms of SBS in order to reach an early diagnosis. The importance of comprehensive history taking and complete physical examination in young children with suspected abusive head injury is vital. The multidisciplinary approach is a key issue in managing such cases in order to rule out medical mimics of abusive head injuries and better safeguard the child. Educational efforts to raise awareness about SBS should be instituted.

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