

Shaken Baby Syndrome: Simple Review Article

Abstract

Shaken Baby Syndrome (SBS) occurs in infants when the head is subjected to excessive acceleration and deceleration. Guthkelch first identified SBS when he noticed that infants with subdural hematoma did not always have gross markings, indicating the possibility of a baby shaking. The rotational force pushes the brain against the skull, causing various types of head and neck injuries. Ophthalmologic testing for retinal haemorrhages and ocular fundus, which can rule out SBS, is one of the tests for SBS. Immunohistochemical staining for β -amyloid precursor protein (β -APP) and magnetic resonance imaging (MRI) accurately identify brain injuries and bleeding, resulting in a more accurate diagnosis of SBS. SBS symptoms are shared by other etiologies, making it difficult to determine the true cause of infantile injury. Experiments using biomechanical models to recreate the whiplash movement have not revealed subdural haemorrhaging, but limitations in the models have doubt to these results.

Key words: Shaken baby syndrome, serious neurological injury, acceleration–deceleration of the head.

Introduction

The most common cause of serious neurological injury or death as a result of child abuse is shaken baby syndrome. It is unique to infancy, when children have distinct anatomical features. Shaking injury is characterised by subdural and retinal haemorrhages. In 1974, an American radiologist named John Caffey coined the term whiplash shaken infant syndrome. Guthkelch, a British neurosurgeon, was the first to identify shaking as the cause of subdural haemorrhage in infants. Impact was thought to play a significant role in the cause of brain damage. Improved neuropathology and imaging techniques have recently established hypoxic ischaemic encephalopathy as the cause of brain injury. The most sensitive and definitive

method of confirming a shaking injury is diffusion weighted magnetic resonance imaging. Social welfare agencies should conduct thorough investigations into the families of children who have subdural haemorrhages. (1)

Shaken baby syndrome is a type of non-accidental physical injury to infants that is characterised by acute encephalopathy with subdural and retinal haemorrhages that occurs in the context of an inappropriate or inconsistent history and is frequently accompanied by other apparently inflicted injuries. Neck and spinal cord injuries are also possible. The precise cause of the brain injury, the retinal and subdural haemorrhages, as well as the degree of force required and whether impact in addition to whiplash forces is required, are all subject to debate. Although the majority of the discussion has focused on fatal injuries of this type, not all are fatal; however, they may be accompanied by subsequent neurological disability of varying severity. (1, 2)

Expert medical evidence about inflicted injury must be scientifically rational, but applying evidence-based clinical practise criteria presents some challenges. Medical management of defined clinical problems can be compared in clinical practise, and best practise can be distinguished by clinical outcomes. In the case of inflicted paediatric injuries, the outcome is presented, the investigation follows rather than precedes the outcome, and the history may be incomplete or intentionally misleading. There is a need for unbiased and intelligent valuation, but how can this be accomplished in practise? Because of the serious implications of diagnosing inflicted injury, such as shaken baby syndrome, each case must be evaluated in detail, taking into account all of the circumstances surrounding the injury and fully considering the pathological features, rather than attempting to assess the significance of each component. (2)

The combined triad of subdural and retinal haemorrhage with brain damage, as well as the characteristics of each of these components, allow a reconstruction of the mechanism of injury and valuation of the degree of force used in shaken baby syndrome. When rotational acceleration and deceleration forces are applied to the infant's head, the brain rotates within the skull. Acute deceleration allows the brain to continue rotating until the connecting veins are

stretched and ruptured, resulting in a thin layer of subdural haemorrhage on the brain's surface. This is not a lesion that takes up space; its significance is in demonstrating the mechanism of injury. The retinal haemorrhages, which are typically extensive, occupy much of the globe's circumference and extend through all layers of the retina, and are caused by rotational acceleration and deceleration forces. (2)

The pathway of brain damage is difficult to understand. According to conventional wisdom, shearing forces operate within the brain substance, causing axonal damage (3). Geddes et al found that the predominant changes in infants with evidence of shaking were hypoxic-ischaemic rather than the diffuse axonal injury seen in older children and adults with fatal head trauma in a careful neuropathological study of head injuries in children using amyloid precursor protein immunostaining (4,5). These authors hypothesised that acceleration and deceleration forces could damage the neuraxis, resulting in apnoea and ischaemic insult, resulting in diffuse cerebral oedema.

Epidemiology

Child abuse is a worldwide issue (27). In Germany, no detailed data on infanticide or the prevalence of SBS are available, either from official registries or from the scientific literature. According to German police statistics for 2006, there are approximately 30 (recorded) cases of abuse and three homicidal deaths per 100 000 children under the age of six. In a multicenter study of SIDS in Germany, autopsies revealed SBS as the cause of death in nearly every 50th case diagnosed as SIDS. The ongoing Survey of Rare Pediatric Diseases in Germany (ESPED, Erhebungseinheit für Seltene Pädiatrische Erkrankungen in Deutschland [in German]) is expected to improve data accuracy even further. Extrapolating the data from the few epidemiological studies—mostly from English-speaking countries, with reporting rates ranging between 15 and 30 per 100 000 children under the age of one year—results in an estimated annual incidence of 100 to 200 cases in Germany. SBS is the most severe form of abuse in infancy and the most common non-natural cause of death; it accounts for more than two-thirds

of all fatal cases of child abuse. Abuse is responsible for more than 90% of all serious intracranial injuries in children. (28)

Risk factors and causes

SBS victims are typically under one year old, rendering them helpless and unable to protect themselves. There are, however, a few case reports of older children with closed head injuries who have severe neurologic impairment and ophthalmic findings that suggest a shaking injury. (6) Because of their relatively large heads and weak cervical musculature, infants and children are especially vulnerable to violent shaking. These factors, along with incompletely fused sutures and relatively large volumes of cerebrospinal fluid in young children, allow for greater movement within the cranial vault, potentially resulting in severe damage to the immature, incompletely myelinated brain. (7) Perceived disruptive behaviour, such as unwarranted and unending crying spells, has been proposed as an important precursor to abuse in the form of 'shaking' by significant figures in the child's life.

SBS perpetrators are typically male, with the biological father being the most common abuser. (8) There is also evidence that stepfathers or male partners may be involved in such heinous acts. Female babysitters, like biological mothers, have been identified as SBS perpetrators. In 72 percent of cases, the perpetrator was a man. (8,9) Males are thought to be more likely to cause SBS when they 'shake' children because of their greater physical power. 1 One could argue that females are more tolerant of their babies' needs and demands. Males, on the other hand, may be more easily provoked by a crying baby. Overall, there is evidence that both parties contribute to this abuse. The distressed child may stress a significant figure, prompting them to abuse the child. [\(9,10\)](#)

Mechanism of injury

SBS is a severe form of abusive head trauma in which a child is held by the torso or the extremities and violently shaken, resulting in rapid head movements with acceleration, deceleration, and rotational forces, with or without impact. It causes a one-of-a-kind combination of intracranial, intraocular, and skeletal injuries. Subdural haematoma is the most

common type of intracranial abnormality found. Retinal haemorrhage is frequently discovered in children with SBS. Fractures of the ribs or long bone fractures when the child is held may also be detected. In cases of SBS, cervical spine injuries are uncommon (6).

Causes and pathophysiology

Child abuse is a multifactorial phenomenon. The risk factors for SBS include low socioeconomic status, disability of the child, violent tendencies, and alcohol or drug abuse within the family. SBS, on the other hand, occurs in all social classes. A typical constellation is a baby who "cries all the time" with young, overworked parents whose repeated attempts to pacify the child fail and who have a low frustration threshold and poor impulse control. In the absence of adequate social resources, a stressful situation may worsen until the child loses control and shakes (29). In surveys conducted in the United States, 50 percent to 75 percent of teenagers and young adults stated that they were unaware that shaking is dangerous, and between 2.6 percent and 4.4 percent of parents of children under the age of two stated that they had shaken their child at least once. In Indian city slums, the equivalent figure was 42 percent. Recently launched public awareness campaigns in the United States have yielded promising early results. These measures, however, have not yet been estimated. It is the courts' responsibility to make the sometimes difficult decision of whether there was any intent to kill or harm the child. Nonetheless, many perpetrators may be well aware of the risks. The perpetrator is usually the child's father or the mother's new partner, but it can also be the mother or a female babysitter. A combination of anatomical features makes infants particularly vulnerable to acceleration-deceleration events with a significant rotatory component, which occur naturally during shaking. The head is large in comparison to the rest of the body, and the weak, immature neck musculature is not yet adequately supporting and controlling it. The result is vigorous movement of various intracranial compartments qualified to one another, such as between the skull and dura on one hand and the cerebral surface on the other, or between the white matter and grey matter. Although many details are still unknown, the vast majority of researchers agree that the resulting shear forces are to blame for subdural haemorrhages and diffuse brain damage. "Simple" shaking without influence is sufficient to

produce the full picture of SBS, with or without fatal outcome, but the energy resulting from an abrupt deceleration through influence is unquestionably greater, leading to more severe trauma (shaken impact syndrome) (28)

Historical review

Child abuse and murder have long been recognised as a phenomenon that has occurred throughout human history. SBS, on the other hand, was not discovered medically until the second half of the twentieth century (20,21,22). In 1946, American paediatrician John Caffey described infants with long bone fractures and subdural haemorrhage (23). Caffey suspected that this constellation was the result of unnoticed or concealed accidents, but he didn't realise that what he was seeing was a typical syndrome following abuse. Henry Kempe published his observations on the "battered child syndrome" in 1962, which was the first comprehensive scientific article on the subject of child abuse. (24). Norman Guthkelch, a British neurosurgeon, described two infants with subdural haemorrhage but no signs of external injury in 1971; the cause, he suspected, was an acceleration-deceleration mechanism ("whiplash injury") (25). Caffey's seminal study on SBS, in which he was the first to link a shaking event with a constellation of subdural haemorrhage, retinal haemorrhage, and long bone fractures, was published in 1972. (26). Caffey is thus regarded as the first to describe SBS, despite the contributions of Kempe, Guthkelch, and others.

Clinical signs

There is a wide range of clinical symptoms. (30,31) The mildest are non-specific, which means that injury may go undetected; the most severe are shocked, unconscious, convulsing children. Even the most inexperienced caregiver will notice that the child is clearly ill immediately following the incident. Poor feeding, vomiting, lethargy, and irritability are non-specific symptoms that can last for days or weeks. Doctors frequently downplay these symptoms, which may be attributed to viral illness, feeding issues, or colic. (32) In some cases, the signs of previous injury may not be recognised until the child is injured again or has a chronic subdural

haematoma (head enlargement). Some children with non-specific symptoms and undetected brain injury are likely to develop learning difficulties and educational failure later in life. It is important to note that, unlike an extradural haemorrhage, there is no lucid interval between the incident and loss of consciousness in the case of a severe injury. (30, 33, and 34). The child may be opisthotonic at presentation, with a full or distended fontanelle. Common symptoms include pallor, hypothermia, and shock. Apnea, irregular breathing, and cyanosis all necessitate intubation and ventilation. Shaken children frequently sustain eye and skeletal injuries.

Diagnosis

SBS is notorious for being difficult to detect and diagnose. Clinicians should use their own clinical judgement because each individual case is unique and must be considered carefully based on the evidence. According to World Health Organization (WHO) estimates, nearly 31,000 children aged 15 died as a result of homicide worldwide in 2002. (11) Despite advancements in investigative neurology, abusive head trauma is frequently misdiagnosed and remains a diagnostic challenge. Any infant or young child who collapses for no apparent reason should be evaluated for SBS. Clinicians must keep a low level of suspicion in mind when considering this diagnosis. (12) SBS is usually diagnosed after a thorough medical and social history is taken. This should be supplemented by appropriate research. Children with SBS are frequently seen first in emergency rooms (EDs). (13)

A thorough history of awarding complaints is an essential component of the diagnostic process. Infants with SBS present to the hospital with a wide range of symptoms, including vomiting, poor suckling, and lethargy, as well as convulsions, apnea, and death. Because symptoms occur immediately after the insult, it is critical to record the timing of the symptoms. Not all infants are critically ill when they present, and in some cases, the lack of a history or external signs of injury may delay diagnosis. It is also worth noting that shaking alone, without any impact injury, can produce the symptoms seen in children with shaken baby syndrome. There are a number of characteristics in children and parents that may raise suspicions of harm; these are triggering influences such as "crying, temperamental behaviour, toileting problems," and a history of previous or recent injury. Birth history, developmental milestones, and vitamin

K status should all be recorded. When a history of injury is provided, it is usually of a minor nature, which is inconsistent with the severity of the infant's condition. A review of risk indicator checklists for child abuse in emergency departments reveals that three history items are worth considering: delay in seeking medical advice, an inconsistent history, and clinical findings that are inconsistent with the history related by the accompanying adult. (6)

SBS physical examinations should cover the entire body, looking for signs of external injuries such as skin bruising, abdominal injuries, and skeletal injuries such as rib or long bone fractures. The child's conscious level, neck and cervical spinal cord injuries, and the need for resuscitation should all be calculated. It is also critical to look for signs of intracranial bleeding, such as fontanelle fullness and increased head circumference. Fundi should be examined by a clinician/paediatrician and, as soon as possible, by an experienced ophthalmologist to rule out eye injury, including retinal haemorrhage. When SBS is suspected, neuro-imaging should be performed as the definitive diagnostic investigation. A computed tomography (CT) head scan, followed by a magnetic resonance imaging (MRI) scan, is the first line of investigation in suspected abusive head trauma.

MRI is a more sensitive method of detecting small intracranial collections, particularly in areas where CT is less effective. (14) Diffusion weighted MRI can also show cerebral oedema and ischaemic changes. A skeletal survey, including skull films, should be performed on all children under the age of three who are suspected of physical abuse (15). The clinical diagnosis is usually based on a patient history that does not describe the clinical features; it is supported by the results of the physical, retinal, and brain MRI examinations.

Differential diagnosis

In total, 95 percent of infants who have suffered a serious intracranial injury have been shaken (35). The remainder are primarily the result of severe head trauma, such as that sustained in a car accident. Other conditions that cause subdural haematomas are the same as those that cause retinal haemorrhage. Small subdural and subarachnoid haemorrhages occur in

approximately 20%–30% of asymptomatic neonates and resolve spontaneously. (1) Birth injury is a rare cause of chronic subdural haemorrhage. (36) Due to the difficulty in obtaining a reliable history, it is unknown how frequently this occurs.(37)

Small amounts of venous bleeding into an enlarged subdural space cause chronic subdural haematomas. This occurs when there is brain atrophy, such as in the rare metabolic disorder glutaric aciduria type 1. (38-39) The enlarged subdural space in this condition is thought to cause stretching of the bridging veins, which tear in response to minor trauma. A vascular membrane with fragile capillaries is formed as a result of granulation tissue ingrowth from the dura. Capillary microhaemorrhages either maintain or enlarge the size of the haematoma. (40) Chronic subdural haematomas should be considered inflicted in all children except those with glutaric aciduria type 1 and ventricular shunts. Frontotemporal atrophy and widening of the sylvian fissure will be visible on brain imaging in glutaric aciduria type 1. A biochemical screen can be used to make the diagnosis, and the absence of glutaryl-CoA dehydrogenase activity in tissue fibroblasts can be used to confirm it. (1)

It has been proposed that infants with macrocephaly and benign enlargement of the subarachnoid spaces are at risk of developing subdural haemorrhage from minor trauma (41). This is in response to reports in the literature of subdural effusions in children with this benign condition. The fact that prominent subarachnoid spaces are common in infants and subdural haemorrhage is uncommon would suggest that there is no scientific basis for that supposition (42) Subdural haemorrhage has been linked to brain shrinkage caused by hypernatraemia-induced cellular water loss. According to extensive research, hypernatraemia is the result of brain injury caused by subdural haematoma rather than the cause. (43)

Management

When abusive head trauma is suspected, a strategy meeting should be held with police and the children's social care provider to determine whether to launch an investigation and then a criminal investigation. Children are frequently referred to a specialised centre with paediatric neuroscience resources. It is critical that such specialists are supported by general

paediatricians who can communicate with local and statutory child protection teams and fully participate in child safeguarding procedures. Laboratory tests are required to rule out other medical conditions such as rare metabolic diseases (glutaric aciduria), coagulation problems, and infective encephalopathy.(16,17)

Other tests should include a septic screen to rule out infection—as subdural collections may be associated with meningitis—urine toxicology screening, and a metabolic screen. It is also critical to perform a full blood count, which should be repeated after 24–48 hours, as this may reveal a rapidly falling and low haemoglobin level. (18). The ophthalmologist's role is frequently to assist in the diagnosis of SBS by examining the child for retinal haemorrhages. Retinal haemorrhages usually resolve on their own and do not require treatment; however, extensive, non-resolving vitreous haemorrhage or retinal detachment may necessitate surgical intervention. A careful follow-up is recommended to document and treat any sequelae that may occur as a result of neurologic or ocular damage.(19)

Prognosis

The prognosis for victims of shaken baby syndrome varies with the severity of injury but generally is poor. Many cases are fatal or lead to severe neurological deficits. Death is usually caused by uncontrollable increased intracranial pressure from cerebral edema, bleeding within the brain or tears in the brain tissue. However, even babies with injuries that appear to be mild may show developmental difficulties. Typically, surviving babies with this syndrome may develop any of the following disabilities:

- Cerebral palsy
- Paralysis
- Vision loss or blindness
- Mental retardation
- Epilepsy
- Seizures

Complications

Even brief shaking of an infant can cause irreversible brain damage. Many children affected by shaken baby syndrome die.

Survivors of shaken baby syndrome may require lifelong medical care for conditions such as:

- Partial or total blindness
- Developmental delays, learning problems or behavior issues
- Intellectual disability
- Seizure disorders
- Cerebral palsy

Conclusion

The shaken baby syndrome sequence of events begins with violent whiplash shaking. Stretch injury to the neuraxis is caused by cervical hyperextension. This causes difficulty breathing or apnoea. Hypoxic ischaemic cerebral injury results from the subsequent hypoxia and shock. Cerebral oedema, intracranial hypertension, and a drop in cerebral perfusion pressure all contribute to additional brain damage. In comparison to traumatic injury in childhood, inflicted injury has a significantly worse prognosis. Subdural and retinal haemorrhage are significant indicators of shaking injury. Diffusion weighted MRI is the quickest, most sensitive, and specific method of determining a shaking injury. This should be done in conjunction with the initial computed tomogram whenever possible. (1)

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