

Shaken Baby Syndrome: Simple Review Article

Abstract

Shaken Baby Syndrome (SBS) occurs in infants when subjected to excessive acceleration–deceleration of the head. SBS was first pointed out by Guthkelch, who noticed that infants with subdural hematoma did not always have gross markings, which signified the shaking of a baby as a possibility. Rotational force pushes the brain against the skull leading to various types of injuries to the head and neck. Examinations for SBS include ophthalmologic testing which tests for retinal hemorrhages and ocular fundus, which may rule out SBS. β -amyloid precursor protein (β -APP) immunohistochemical staining and magnetic resonance imaging (MRI) accurately identify brain injuries and bleeding leading to a more accurate diagnosis of SBS symptoms of SBS are shared by other etiologies, making it difficult to distinguish the true source of infantile injury. Experiments that have attempted to recreate the whiplash movement with biomechanical models have not shown subdural hemorrhaging, but limitations in the models cast doubt to these results. Shaken Baby Syndrome (SBS) occurs in infants when subjected to excessive acceleration–deceleration of the head SBS was first pointed out by Guthkelch, who noticed that infants with subdural hematoma did not always have gross markings, which signifies the shaking of a baby as a possibility. Rotational force pushes the brain against the skull leading to various types of injuries to the head and neck. Examinations for SBS include ophthalmologic testing which tests for retinal hemorrhages and ocular fundus, which may rule out SBS. β -amyloid precursor protein (β -APP) immunohistochemical staining and magnetic resonance imaging (MRI) accurately identify brain injuries and bleeding leading to a more accurate diagnosis of SBS. Symptoms of SBS are shared by other etiologies, making it difficult to distinguish the true source of infantile injury. Experiments that have attempted to recreate the whiplash movement with biomechanical models have not shown subdural hemorrhaging, but limitations in the models cast doubt to these results.

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

Introduction

Shaken baby syndrome is the most common cause of serious neurological injury or death resulting from child abuse. It is specific to infancy, when children have unique anatomic

features. Subdural and retinal haemorrhages are markers of shaking injury. An American radiologist, John Caffey, coined the name whiplash shaken infant syndrome in 1974. It was, however, a British neurosurgeon, Guthkelch who first pronounced shaking as the cause of subdural haemorrhage in infants. Impact was advanced thought to play a major part in the causation of brain damage. Lately improved neuropathology and imaging techniques have established the cause of brain injury as hypoxic ischaemic encephalopathy. Diffusion weighted magnetic resonance imaging is the most sensitive and definite method of confirming a shaking injury. Families of children with subdural haemorrhages should be carefully investigated by social welfare agencies(1)

Shaken baby syndrome is a form of physical non-accidental injury to infants, characterised by acute encephalopathy with subdural and retinal haemorrhages, happening in a context of inappropriate or inconsistent history and commonly accompanied by other apparently inflicted injuries. Injuries to the neck and spinal cord may also be existing. Controversy surrounds the precise causation 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 needed. Although most discussion has concerned fatal injuries of this nature, not all are lethal, but they may be accompanying with subsequent neurological disability of varying severity.

Expert medical evidence about inflicted injury must have scientific rationality, but applying the evidence based criteria appropriate to clinical practice entails some difficulties. In clinical practice medical management of defined clinical problems can be compared and best practice distinguished by clinical results. Conversely, in inflicted paediatric injuries, one is presented with the outcome, investigation monitors rather than precedes that outcome, and the history may be incomplete or deliberately misleading. A need exists for an impartial and intelligent valuation, but how may this be achieved in practice? Because of the serious implications of diagnosing inflicted injury such as shaken baby syndrome, every case must be evaluated in detail, taking account of all the circumstances adjacent the injury and considering

the pathological features in full, rather than attempting to evaluate the significance of each component.(2)

In shaken baby syndrome, it is the combined triad of subdural and retinal haemorrhage with brain damage, as well as the features of each of these components that allow a reconstruction of the mechanism of injury, and valuation of the degree of force employed. The application of rotational acceleration and deceleration forces to the infant's head causes the brain to rotate in the skull. Abrupt deceleration allows continuing brain rotation until connecting veins are stretched and ruptured, causing a thin layer of subdural haemorrhage on the surface of the brain. This is not a space occupying lesion; its importance is in demonstrating the mechanism of injury. The retinal haemorrhages, which are characteristically extensive, occupy much of the circumference of the globe and extend through all the layers of the retina and similarly result from rotational acceleration and deceleration forces.(2)

The mechanism of brain damage is problematic. Traditional wisdom has suggested shearing forces operating within the brain substance with consequent axonal damage(3). Geddes et al, in a careful neuropathological study of head injuries in children using β amyloid precursor protein immunostaining, observed 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(4,5). These authors thought that acceleration and deceleration forces might damage the neuraxis to cause apnoea, with consequent ischaemic insult causing diffuse cerebral oedema.

Epidemiology

Child abuse is a global problem (27). No detailed data on the killing of infants or on the incidence of SBS are available for Germany, neither from official registries nor in the scientific literature. Analysis of German police statistics for the year 2006 indicates an annual incidence of around 30 (recorded) cases of abuse and three homicidal deaths per 100 000 children under the age of 6 years . In a multicenter study of sudden infant death (SIDS) in Germany, autopsy revealed SBS as the cause of death in almost every 50th case diagnosed as SIDS. Further improvement in the accuracy of the data can be expected from the ongoing Survey of Rare

Pediatric Diseases in Germany (ESPED, Erhebungseinheit für Seltene Pädiatrische Erkrankungen in Deutschland [in German]). Extrapolation of the data from the few epidemiological studies—principally from English-speaking countries and reporting rates of between 15 and 30 per 100 000 children under the age of 1 year—yields an estimated incidence of 100 to 200 cases per year in Germany. SBS is the most severe form of abuse in infancy and the most frequent non-natural cause of death; more than two-thirds of all fatal cases of child abuse occur in this age group. Over 90% of all serious intracranial injuries in infancy result from abuse (28)

Risk factors and causes

The victims of SBS are usually under one year-old and thus helpless and unable to protect themselves. 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.⁽⁶⁾ Infants and children are particularly vulnerable to violent shaking because of their relatively large head and weak cervical musculature. 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.⁽⁷⁾ 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.

Perpetrators of SBS are often male with the biological father being the most common abuser.⁽⁸⁾ 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.^(8,9) 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(9,10)

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. The most common intracranial abnormality detected is subdural haematoma. Children with SBS are often found to have retinal haemorrhage. 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(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. However, SBS occurs in all social strata . A typical constellation is that of a baby that "cries all the time" with young, overstressed parents whose repeated efforts to pacify the child end in failure and who have a low frustration threshold and poor control of impulses. In the case of inadequate social resources, a stress situation may grow ever more acute until it ends with loss of control and shaking of the child (29). In surveys carried out in the USA, 50% to 75% of teenagers and young adults detailed they did not know that shaking is dangerous and between 2.6% and 4.4% of the parents of children under 2 years of age described they had shaken their child at least once . The equivalent figure for parents in Indian city slums was 42% . Recently initiated public awareness campaigns in the USA have shown auspicious early results. Efforts to raise consciousness of the particular dangers of shaking and of the help available have also begun in Germany, exactly in Hamburg and Lower Saxony (leaflet for young parents in Hamburg: "Hilfe! Mein Baby hört nicht auf zu schreien," www.hag-gesundheit.de/documents/schuette_130.pdf [in German]). However, these measures have not

yet been estimated. It is the responsibility of the courts to make the sometimes particularly difficult judgment of whether there was any meaning to kill or harm the child. Nevertheless, many perpetrators are maybe well aware of the dangers .The perpetrator is most commonly the child's father or the mother's new partner, less frequently the mother or a female babysitter. A number of anatomical features make infants mainly vulnerable to acceleration-deceleration events with a marked rotatory component, which naturally occur on shaking . The head is large in relation to the rest of the body and is not yet adequately supported and controlled by the weak, immature neck musculature . The effect is vigorous movements of the various intracranial compartments qualified to one another, e.g., between the skull and dura on the one hand and the cerebral surface on the other, or between the white matter and the gray matter. Although many details remain unclear, the overwhelming majority of investigators agree that the resulting shear forces are responsible for subdural hemorrhages and diffuse brain damage . "Simple" shaking without influence suffices to produce the full picture of SBS with or without fatal outcome, but the energy resulting from an abrupt deceleration through influence is certainly higher and thus leads to more severe trauma (shaken impact syndrome)(28)

Historical review

Abuse and killing of children has long been recognized as a phenomenon occurring throughout human history. However, the medical discovery of SBS did not begin until the second half of the 20th century (20,21,22). In 1946 the American pediatrician John Caffey described infants with fractures of the long bones and subdural hemorrhage (23). Caffey suspected that this constellation could have arisen from unnoticed or concealed accidents, but did not yet realize that what he was seeing was actually a characteristic syndrome following abuse. In 1962 Henry Kempe published his observations on the "battered child syndrome," the first comprehensive scientific article on the topic of child abuse (24). In 1971 the British neurosurgeon Norman Guthkelch described two infants with subdural hemorrhage but no signs of external injury; as the cause, he suspected an acceleration-deceleration mechanism ("whiplash injury") (25). In 1972 followed Caffey's seminal study on SBS, in which he was the

first to link a shaking event with a constellation, henceforth recognized as typical, of subdural hemorrhage, retinal hemorrhage, and fractures of the long bones (26). Caffey is therefore regarded—while acknowledging the important work done by Kempe, Guthkelch, and others—as the first to describe SBS.

Clinical signs

There is a wide spectrum of clinical signs.(30,31) The mildest are non-specific so that injury may never be detected; the most severe being the shocked, unconscious, convulsing child. Immediately after the incident the child will always be obviously unwell, even to the most inexperienced carer. The non-specific signs that may persist for days or weeks are poor feeding, vomiting, lethargy, and irritability. These signs are often minimised by doctors and may be attributed to viral illness, feeding problems, or colic.(32) In some, the signs of previous injury may only be recognised when the child is injured again or presents with a chronic subdural haematoma (head enlargement). It is likely that some children with non-specific signs and undetected brain injury present later with learning difficulty and educational failure.

In the case of a severe injury, it is important to note that unlike an extradural haemorrhage there is no lucid interval between the incident and loss of consciousness.(30, 33,34). At presentation the child may be opisthotonic with a full or distended fontanelle. Pallor, hypothermia, and shock are common signs. Apnoea, irregular respiration, and cyanosis necessitate intubation and ventilation.

Children who have been shaken frequently have eye and skeletal injuries.

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.(11) 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.(12) 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.(13)

A comprehensive history of the awarding complaints is an vital component of the diagnostic process. Infants with SBS present to hospital with a variation of symptoms ranging from vomiting, poor suckling and lethargy to convulsions, apnoea and death. Symptoms happen immediately after the insult, thus recording the timing of the symptoms is very important. Not all infants are intensely ill at presentation, and in some cases the absence of either a history or external signs of injury may delay diagnosis. It is also significant to note that shaking alone without impact injury can produce the indications seen in children with shaken baby syndrome. There are a quantity of features in the children and parents that may increase suspicion that harm has been done; 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 are also important to note. In most cases where a history of injury is given, it is stated 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 related by the accompanying adult.(6)

Physical examinations for SBS should contain 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. The child conscious level, neck and cervical spinal cord injuries should be estimated together with the need for resuscitation. It is also essential 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 conceivable by an experienced ophthalmologist, in order to exclude eye injury including retinal haemorrhage. Neuro-imaging is the definitive diagnostic investigation and should be performed when 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 an additional sensitive method of detecting small intracranial collections, especially in areas less well seen on CT.(14) 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(15). The clinical diagnosis is usually based on a patient history that does not describe the clinical features; it is supported by the findings of the physical and retinal examinations and the brain MRI.

Differential diagnosis

Altogether 95% of infants with a serious intracranial injury have been shaken(35). The remainder are mostly caused by severe head trauma as in motor vehicle accidents. Other conditions that cause subdural haematomas are the same as those which have been listed as causing retinal haemorrhage. About 20%–30% of asymptomatic neonates have small subdural and subarachnoid haemorrhages which rapidly resolve spontaneously.(1) Rarely, chronic subdural haemorrhage is caused by birth injury.(36) How often this happens is unknown owing to the difficulty in obtaining a reliable history.(37)

Chronic subdural haematomas develop from small amounts of venous bleeding into an enlarged subdural space. This arises where there is brain atrophy such as in the rare metabolic condition glutaric aciduria type 1.(38-39) It is thought that in this condition the enlarged subdural space causes stretching of the bridging veins which tear in response to minimal trauma. An ingrowth of granulation tissue from the dura then forms a vascular membrane with fragile capillaries. Capillary microhaemorrhages maintain the size or enlarge the haematoma.(40) Other than in children with glutaric aciduria type 1 and ventricular shunts,

chronic subdural haematomas should be regarded as inflicted. In glutaric aciduria type 1 frontotemporal atrophy and widening of the sylvian fissure will be apparent on brain imaging. The diagnosis can be made on a biochemical screen and confirmation obtained by the absence of glutaryl-CoA dehydrogenase activity in tissue fibroblasts.(1)

It has been suggested that infants with macrocephaly and benign enlargement of the subarachnoid spaces are at risk of developing subdural haemorrhage from minor trauma(41). This follows 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 rare, would indicate that there is no scientific basis for that supposition(42)Brain shrinkage, from cellular loss of water in hypernatraemia, has been linked with subdural haemorrhage. An extensive investigation has suggested that hypernatraemia is the consequence of brain injury accompanying subdural haematoma and not the cause.(43)

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.(16,17)

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.(18). 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.(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 sequence of events in shaken baby syndrome is initiated by violent whiplash shaking. Cervical hyperextension causes stretch injury to the neuraxis. This produces breathing difficulty or apnoea. The ensuing hypoxia and shock cause hypoxic ischaemic cerebral injury. Further brain damage occurs as a consequence of cerebral oedema, intracranial hypertension, and a fall in cerebral perfusion pressure. In comparison with traumatic injury in infancy, inflicted injury has a much worse prognosis. Subdural and retinal haemorrhage are important markers of shaking injury. The fastest, most sensitive and specific method of determining a shaking injury is diffusion weighted MRI. Where possible this should be undertaken in conjunction with the initial computed tomogram (1)

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