What Does The Recent Literature Tell Us About Shaken Baby Syndrome?

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Background:

The term Shaken Baby Syndrome (SBS) refers to the various signs, symptoms, clinical, radiographic and, in fatal cases, autopsy findings resulting from violent shaking and/or impacting the head of an infant or young child. Some have suggested the alternative term “Shaken Impact Syndrome (SIS)” since in many cases impact is demonstrable and is thought to be responsible for some of the traumatic lesions seen. During such assaults, the bridging veins running from the surface of the brain to the superior sagittal sinus, rupture and bleed into the subdural and/or subarachnoid spaces. Direct traumatic damage occurs to the brain; hypoxia during and after the assault causes further irreversible damage to brain tissue; and the cascade of injury continues as there is breakdown of dying brain cells that release intra-cellular enzymes, injuring adjacent neurons. The combined effect is destruction of brain tissue, leading to cerebral edema, raised intra-cranial pressure within the closed skull of the infant, decreased blood flow to the brain and a vicious circle of anoxia, cerebral edema, and death of brain tissue. These insults to the brain cause the signs, symptoms, radiologic and laboratory findings that characterize the course of this form of inflicted brain injury. The signs and symptoms seen are mild to severe, on a continuum from a “low-dose” of shaking/impact to a “high-dose” of shaking/impact and severe craniocerebral injury and may run the gamut from decreased responsiveness, poor feeding, irritability, lethargy and hypotonia to convulsions, vomiting, tachypnea, hypothermia, bradycardia, coma, fixed dilated pupils to death.

Associated injuries may also be seen outside of the brain itself. Retinal hemorrhages that are numerous, prominent, multilayered, and seen throughout the retina, extending forward to the edge of the retina, are present in the majority but not all cases. The presence of acute skull fractures indicates that impact has accompanied the shaking. Posterior rib fractures are present in less than a third of cases, and are the result of the levering of the arcs of the ribs across the transverse processes of the vertebral bodies during the shaking process. Long bone fractures, especially the classic metaphyseal fracture at the ends of long bone, and skin bruises, particularly of the face, head, and extremities may also occur. Injuries to the bones, interspinous ligaments and muscles of the neck have been described but the true incidence of these lesions is unknown. Nearly a quarter of the victims die within a few hours to a few days of their brain injury. From half to ninety-percent of the survivors are left with varying degrees of disabilities ranging from serious learning disabilities and behavioral disorders to paralyses, blindness, and permanent vegetative states.

Recent literature supports this view of inflicted head injury due to shaking with or without impact. A recent article from the Royal College of Ophthalmologists in the United Kingdom [1] reinforced the belief that normal handling of infants was “highly unlikely” to produce retinal hemorrhages and intracranial bleeding in children. They also asserted that retinal hemorrhages are not caused by cervical injuries alone. Hypoxia resulting from transient apnea does not result in the SBS picture, although hypoxia can cause cerebral edema. They also reaffirmed that short falls

do not cause the retina picture of shaken baby syndrome. Finally, they asserted that intracranial bleeding is not the cause of retinal hemorrhages.

Another article [2] looked at 75 cases of shaken baby syndrome and found that 85% had retinal hemorrhages; 81% were bilateral; 82% were confluent, multiple, and multilayered; and traumatic retinoschisis was present in one third. There was no association between the side of the intracranial bleeding and the retinal bleeding; and there was no correlation between the presence of impact and the presence of retinal hemorrhages. The 29 patients with raised intracranial pressure did not differ in type, extent or frequency of retinal findings compared with the rest of the patients. No correlation was found between the side of the intracranial bleeding and the side of the retinal hemorrhages, thus not supporting the theory of direct tracking of blood through the optic nerve into the retina.

A new technique for identifying possible victims of inflicted traumatic brain injury was recently described by Berger and her colleagues at the University of Pittsburgh. [3] Three chemical substances found in serum and/or cerebrospinal fluid [(neuron-specific enolase (NSE), S100B and myelin-basic protein (MBP)] were studied to determine whether they were sensitive and specific for for inflicted traumatic brain injury (iTBI) in high risk infants. They recruited 98 infants under the age of 1 year with one of the following complaints: apparent life-threatening event; more than 4 episodes of vomiting and no diarrhea within the previous 24 hours; seizure; or other non-specific neurologic sign or symptom such as lethargy or fussiness, symptoms that have been linked to possible iTBI. In addition to the standard workup for these conditions, the authors obtained samples of blood and cerebrospinal fluid and these were analyzed for NSE, S100B and MBP. NSE was 76% sensitive and 66% specific and MBP was 36% sensitive and 100% specific for inflicted traumatic brain injury. S100B was increased in 90% of patients with no brain injury and therefore was not specific for inflicted traumatic brain injury in this population. The authors conclude that their results suggest that serum and cerebrospinal fluid concentrations of NSE and MBP have the potential to be used as screening tests for inflicted traumatic brain injury in well-appearing infants who present without a history of trauma and with non-specific symptoms such as vomiting and fussiness. Seventy-nine percent (11 of 14) of the infants with inflicted traumatic brain injury in this study had an increased NSE or MBP concentration. None had a history of trauma. All appeared well to the examining physician and had presented with non-specific symptoms that could easily be misdiagnosed as other more common conditions, such as colic or gastro-esophageal reflux. These increases in serum concentrations for NSE and MBP might be useful as a screening test to identify infants who would benefit from additional evaluation with a head CT. These tests, however, are not intended as a diagnosis for SBS.

Starling and her associates analyzed the confessions of 81 perpetrators of inflicted head trauma and compared these cases to 90 cases where there was no admission. [4] In the group of 81 confessed cases, all had intracranial bleeding, 83% had retinal hemorrhages, 15% had other abuse-related injuries, 19% of the patients died from their injuries acutely, 71% of the survivors were impaired and only 10% of the survivors were considered normal at discharge. The commonest confessed perpetrator was the father in 56% of the cases, followed by mother's boyfriend in 16%, the mother in 15%, a babysitter in 5% and "other persons" in 9%. The commonest initial symptoms reported by the perpetrators were limpness in (29), seizures (25), vomiting (24), lethargy (23) and apnea (21). Seizures and apnea were the commonest symptoms noted by health care personnel at the time of admission, and there were no significant differences between what the perpetrators and the medical staff described. In 91% of

the 57 cases where a time interval between injury and the appearance of symptoms could be determined, the perpetrators said the symptoms appeared “immediately” after the injury.

Keenan and her colleagues compared 80 cases of inflicted traumatic brain injury to 72 with non-inflicted injury. [5] Sixty-four percent of the inflicted injury group had no explanation for the head injury. None of the non-inflicted group had no explanation. The reasons for seeking care in the inflicted group were breathing difficulty in one third, lifelessness, and seizures, but these symptoms were not prominent in the non-inflicted group where lethargy and irritability were the most prominent symptoms. Retinal hemorrhages were present in 76% of the inflicted group but only in 8% of the non-inflicted group. Those in the inflicted group were more likely to have Glasgow Coma Scores between 9 and 12, had longer hospital stays and had poorer outcome scores.

Publications about shaken baby syndrome are more numerous than ever before. Despite the challenges in studying this condition, there are hundreds of papers on SBS from many countries in the world. The evidence continues to build on past concepts and the studies that are being published are increasingly convincing that earlier clinical observations and judgments were not in serious error. As Minns has stated, “the lack of an evidence base…does not negate the syndrome: it just means the support for the syndrome is incomplete.”[6]

1. Adams G et al: Update: Ophthalmology working party. Eye 2004;18:795-798
2. Morad Y et al; Correlation between retinal abnormalities and intracranial abnormalities in shaken baby syndrome. Amer J Ophthalmol 2002;134:354-359)
3. Berger RP, Dulani T, Adelson D, Leventhal JM, Richichi R, Kochanek P. Identification of inflicted traumatic brain injury in well-appearing infants using serum and cerebrospinal markers: a possible screening tool. Pediatrics 2006;117;325-332.
4. Starling SP et al. Analysis of perpetrator admissions to inflicted traumatic brain injury in children. Arch Pediatr Adolesc Med 2004;158:454-
5. Keenan HT, Runyan DK, Marshall SW, Nocera MA, and Merten DF. A population-based comparison of clinical and outcome characteristics of young children with serious inflicted and non-inflicted traumatic brain injury. Pediatrics 2004;114:633-639.
6. Minns R. Shaken baby syndrome: theoretical and evidential controversies. J Royal Coll Physicians Edinburgh 2005;35:5-15.