

# New Developments in 'Shaken Baby Syndrome'

Inflicted head injury is one of the commonest causes of explained death in infancy.<sup>1</sup> Following a number of high profile cases including an Appeal Court judgement, there has been increasing legal and medical controversy surrounding the diagnosis of inflicted head injury in children and in particular the triad of pathological findings that define the 'Shaken Baby Syndrome' (SBS).<sup>2,3</sup> We will discuss the basis for this diagnosis and some of the hypotheses that are offered to explain the pathophysiology of this syndrome.

## Inflicted head injury in infancy

Inflicted head injury in infancy is associated with a distinct pattern of injury characterised by a triad of retinal haemorrhage, sub-dural haemorrhage and an encephalopathy in the absence of any other explanation<sup>1,4</sup> (Table 1). Each of these features has specific characteristics that suggest the diagnosis but it is the combination of the features in the absence of an adequate explanation that carries the most diagnostic significance.

**Table 1: The triad of inflicted head injury in infancy**

- |                                    |
|------------------------------------|
| 1. Retinal haemorrhage             |
| 2. Thin film sub-dural haemorrhage |
| 3. Encephalopathy                  |

In fatal cases, the retinal haemorrhage is typically extensive, bilateral and affects multiple layers of the retina.<sup>5</sup> The nature of the sub-dural haemorrhage differs from those typically seen in adults. In infants, including those with fatal injuries, the sub-dural blood tends to be a low volume 'thin film' and is frequently bilateral.<sup>6</sup> In fact, it is unusual for the sub-dural haemorrhage to exert mass effect and its diagnostic significance is as an indicator of the mechanism of injury rather than the direct cause of death.

The pattern of injury seen within the brain also differs from that seen in older individuals. While a number of typical features of trauma may be seen in infants with inflicted head injury such as gliding contusions of the white matter (which are typically parasagittal contusions associated with acceleration-deceleration injuries), in most cases the pattern of injury is predominantly that of cerebral oedema and hypoxic-ischaemic damage.<sup>7</sup> The widespread pattern of diffuse traumatic axonal injury that is typical of severe closed head injuries in adults is more unusual in inflicted head injury in infants and traumatic axonal injury is often limited to the cortico-spinal tracts around the junc-

tion between the cervical spinal cord and medulla.

While a number of clinical syndromes have been associated with abusive head injury in children,<sup>8</sup> in fatal cases a commonly reported clinical history is that of a previously well infant that undergoes a sudden collapse in the care of a lone adult without corroborative witnesses. In a number of well-documented cases, the carer has confessed to a loss of temper followed by shaking the child with varying degrees of vigour.<sup>9</sup>

While in many cases the pathological findings are limited to the triad described above, in a proportion of cases there is additional evidence of non-accidental injury lending further support to the view that the injuries seen in the triad are due to inflicted trauma.<sup>6</sup>

## Why does head injury in infants show a distinct pathology?

Two factors are likely to explain the pattern of head injury seen in infants. The first is that the infant head and neck have unique mechanical properties that influence their response to trauma (Figure 1). The second is that the nature of the injury is likely to be different in older children and adults.

In infants, the head is relatively heavy and the neck musculature relatively weak. In some cases, there is evidence of traumatic axonal injury at the cervico-medullary junction and this would support the view that the cervico-medullary junction is vulnerable in infants.

The cranial sutures have not fused at birth, which means that the cranial cavity is not the closed box system of the adult and therefore changes in intracranial pressure will behave less predictably. In addition, following an injury one would expect the bones to mould with respect to each other, increasing the torsion on underlying structures, including the dural veins and sinuses. In addition, the base of skull is smoother in infancy affecting injury to the basal structures (e.g. contusions, cranial nerve injuries).

Finally, at birth the brain is poorly myelinated, leading to a more fluid consistency and to a reduced difference in inertia between the grey and white matter. These factors can clearly alter the response of the brain to acceleration-deceleration injuries and may explain the relative rarity of typical traumatic axonal injury in infancy outside the cervico-medullary junction.

The second contributor to the pattern of pathology in infancy is that the nature of the traumatic insult differs. Transcripts of perpetrator's confessions support the assertion that shaking of a child with the head unsupported leads to the typical pattern of injury.<sup>9</sup> In contrast shaking is



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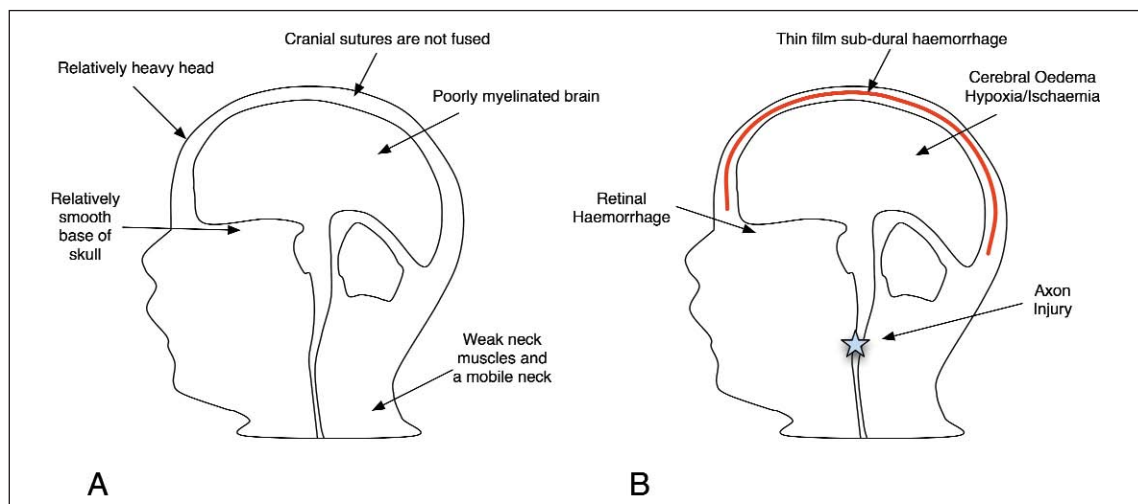
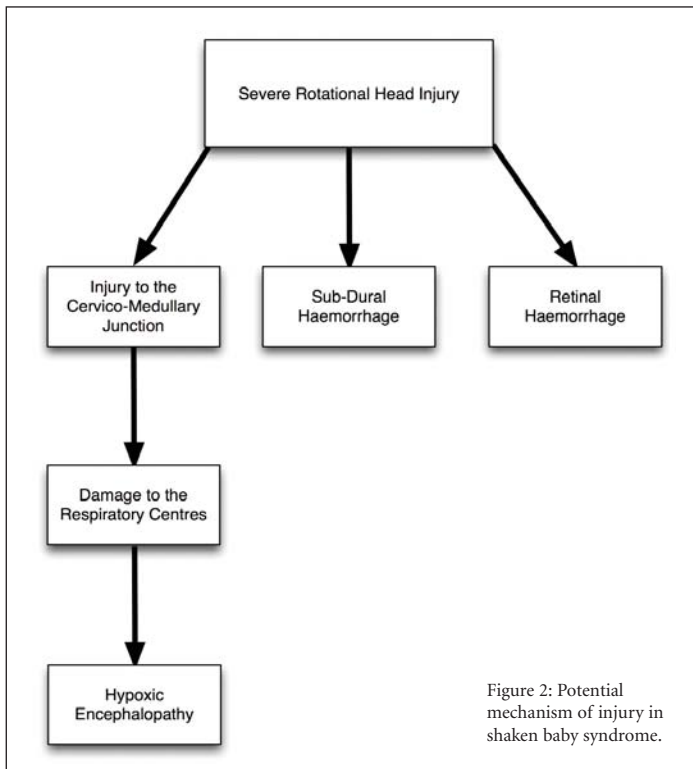


Figure 1: A. The infant brain has unique mechanical features that underlie the pattern of injury seen in inflicted head injury. The figure highlights potential sources of vulnerability. B. The typical pattern of injury in inflicted head injury in infancy.



an unusual cause of death in older children where road traffic accidents and other impact injuries predominate. Controversy surrounds whether shaking is sufficient alone or if shaking in combination with an impact is necessary (e.g. 10). However, in many cases there is no evidence of an impact and, more importantly, the pattern of injury is not that seen in impact injuries.

A plausible hypothesis is that shaking an infant leads to stretching of the cervico-medullary junction due to the particular mechanical vulnerability at this site in infants. This injury causes a respiratory disturbance or apnoea through damage to the respiratory centres and their connections. Finally, the respiratory failure causes a global hypoxic injury to the brain. In such a model, the concurrent retinal and sub-dural haemorrhages are simply indicators of a severe rotational injury (Figure 2).

### Controversy in the shaken baby syndrome

Some authors have proposed that the pattern of injury labelled Shaken Baby Syndrome may be caused by non-violent mechanisms.<sup>3</sup> For example, as an extension of the observation that hypoxic / ischaemic brain injury is common in the triad, some authors have asked can hypoxia or ischaemia cause all the features of shaken baby syndrome? We believe this is unlikely as there is no well-documented clinical evidence of the syndrome arising in the presence of a defined hypoxic / ischaemic insult. Furthermore, such a hypothesis fails to explain the evidence of confessions, the cases in which there are extra-cranial injuries and the cases in which there is other evidence of traumatic head injury (e.g. gliding contusions, skull fractures etc or even traumatic axonal injury in the medulla).

In one small cohort of paediatric autopsies, fresh intra-dural haemorrhage was a relatively frequent finding at post-mortem in fetal, perinatal and infant autopsies in the absence of evidence of trauma.<sup>11</sup> However, macroscopic sub-dural haemorrhage is the important finding in SBS, not microscopic intra-dural haemorrhage. Indeed, the authors did not see macroscopic thin film sub-dural haemorrhage (with or without the rest of the triad) in their non-trauma infant cohort. Furthermore, the cohort contained only nine infants, the remainder of the cases being made up with perinatal and fetal cases. In addition, the retina was not examined in these cases. While the cohort included frequent evidence of hypoxic-ischaemic injury, the authors did not show a statistically significant correlation between hypoxia and intra-dural haemorrhage.

A further explanation proposed for the sub-dural bleeding is that it arises during birth or that a sub-dural haematoma that formed at birth has re-bled. If re-bleeding from these haematomas were the cause of death, one would predict that it would need to generate a space-occupy-

ing lesion with mass effect. Such a space-occupying mass is not seen in the triad and therefore it seems unlikely that re-bleeding is a mechanism of death. Furthermore, in a careful longitudinal study, congenital sub-dural haematomas had a different distribution to that seen in SBS and importantly they resolve by the age of four weeks.<sup>12</sup>

An alternative hypothesis that has been proposed is that paroxysms of coughing, possibly associated with feeding difficulties or choking, may generate a massive rise in intracranial venous pressure that leads to sub-dural and retinal haemorrhage and may be associated with venous infarction.<sup>13</sup> However, there is no well-documented direct evidence that this occurs and it does not explain the cases in which there is direct evidence of trauma. The hypothesis has been based on a mathematical model that estimates the rise in venous pressure during a paroxysm of coughing. In this model, rapid rises in venous pressure are predicted and the authors have argued that there are areas of vulnerability in the veins bridging the sub-dural space that may rupture in the face of a sudden rise in venous pressure. However, the site of bleeding in SBS is not known with any certainty and the pressure needed to rupture the veins in human infants is not known. As with all biomechanical models, the hypothesis generated would have to be validated with good clinical data.

### Conclusions

A general issue at the heart of the debate around SBS is that the quality of scientific evidence that informs it has serious limitations. Central to this is the clinical evidence. However, even in the carefully documented cohort of Geddes, there are only 37 cases.<sup>6,7</sup> Cases with corroborative evidence of trauma (e.g. fractures) are persuasive of the association with inflicted injury but the events are not witnessed and therefore the evidence from legal confession is also an important part of the case for a traumatic mechanism. A number of experimental approaches have been undertaken such as biomechanical models, animal models and cadaveric studies but each of these has been beset with controversy and difficulties in validating the unique mechanics of the infant head and neck. Future studies must include unbiased cohorts of post-mortem studies with standardised protocols of suspicious cases and importantly of non-suspicious cases. It is unfortunate that recent changes in regulation in the UK have made these sorts of studies increasingly difficult. The understandable and justifiable emphasis on parental consent for tissue research taken together with the increasing pressures on the coronial system are likely to introduce significant bias into future cohort studies in the UK.

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