Inflicted Childhood Neurotrauma (Shaken Baby Syndrome)

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n a child presenting with multiple injuries, subdural haematoma and retinal haemorrhages where there is no underlying systemic cause and no evidence of severe accidental trauma, non-accidental injury is considered to be the likely cause; this is the case even when the method of injury is unknown and in the absence of an appropriate animal or mathematical model because observational studies overwhelmingly support this conclusion. In practise the diagnosis is made upon the history, medical evidence from paediatricians, radiologists, ophthalmologists, and forensic examination and reconstruction of the scene and events by the prosecuting authorities.

Previous evidence has suggested that the shaking of a relatively heavy head about the neck causes such inertial forces within the brain tissue that shearing takes place with rupture of meningeal vessels and diffuse axonal injury, resulting in subdural haemorrhage and neurological damage. The retinal haemorrhages are generally thought to be due to the same shearing mechanisms at work within the vitreoretinal interface. Green et al. found a correlation between intra-ocular bleeding, anterior optic nerve haemorrhage and subdural haematomas. When looking at the relative positions of subhyloid haemorrhages at post mortem, it was found that the frequency of position of these haemorrhages coincided with the areas of maximal vitreoretinal adhesion, that is the ora serrata and the optic disc. They hypothesised ‘...the mass of the vitreous acts upon the points at which it is attached during both acceleration or deceleration...producing first small haemorrhages in the retina then retinal detachment...’ Post mortem findings of vitreous traction at the apex of retinal folds and the edge of dome shaped haemorrhages and retinoschisis gives some supporting evidence that vitreous forces may cause this shearing damage. There is no adequate model to test this experimentally, so this remains hypothesis, not established fact. In the situation of isolated intraocular haemorrhage with or without accompanying subdural haemorrhage there continues to be disagreement regarding the possibility of an accidental injury such as a short fall in the domestic setting being responsible for the clinical findings in the absence of other evidence to support non-accidental injury.

The Royal College of Ophthalmologists Working Party concluded in terms of the force required to cause retinal haemorrhages ‘...no absolute values can be given for the angular acceleration forces required to produce...injury, but there is good evidence that they must be considerable...’

In 2003 ‘Brain haemorrhage in babies may not indicate violent abuse’ appeared as a headline in the BMJ following a not proven verdict in the case of a child minder accused of murder at the High Court in Edinburgh. During this case research by Geddes et al. was presented which purportedly cast doubt that the injuries could only have been due to violent shaking.

Geddes and colleagues in a series of papers presented their findings that the most common pathological finding was of brain swelling and hypoxic ischaemic encephalopathy with a significant number of cases having focal axonal damage in the lower brainstem rather than diffuse axonal injury as previously thought. They hypothesized that damage to the brainstem, by hyperextension–flexion (shaking) injury at the cranio-cervical junction may cause focal damage, resulting in apnoea, and a cascade leading to hypoxic ischaemic encephalopathy, brain swelling, raised intracranial pressure and death. (This has been referred to as the ‘unified hypothesis’.) They further suggested that hypoxia-related leakage of blood from veins both inside the dura and in the subdural space was the source of the subdural haemorrhage rather than traumatic rupture of bridging veins and that in the immature brain hypoxia alone is sufficient to activate the pathophysiological cascade which culminates in altered vascular permeability and extravasation of blood, so that the subdural and retinal haemorrhages were a secondary phenomenon and not due to shearing forces. This led to the conclusion that ‘...it may not be necessary to shake an infant very violently to produce stretch injury to the neuroaxis...’ These comments related to those difficult cases in which there was little or no external evidence of injury but there were retinal and thin film subdural haemorrhages.

Other researchers have similarly reported the finding of hypoxic ischaemic damage rather than diffuse axonal injury but have not drawn the same conclusions regarding the forces involved.

However in terms of ocular examination there is no description of the retinal haemorrhages, they were either present or absent and they do not mention whether the optic nerves were examined. In the first paper they comment that ‘...a discussion of the aetiology of retinal haemorrhages...is beyond the scope of this paper. Later in their third paper they discuss the possible cause of retinal haemorrhages; ‘...retinal haemorrhages can be explained by rises in intracranial pressure and central venous pressure, with and without hypoxia; they are also seen in a proportion of normal infants at birth, as well as in premature babies. In the setting of inflicted infant head injury, it has never been proved that retinal bleeding is directly caused by
shaking; rather, it is widely assumed that it results from the shearing forces of the injury, which simultaneously cause retinal and subdural bleeding and diffuse brain damage. However...most infant victims...show very little ...traumatic pathology in the brain, it is appropriate to re-evaluate this assumption..."

Geddes pathological findings add little new evidence to the knowledge of retinal haemorrhages in shaken baby syndrome. In evaluating the latter Professor Luther9 an ophthalmic pathologist reviewing the various theories of causation (shearing forces vs other rheological mechanisms), timing and nature of injury concluded”….I consider it premature to...”

The Geddes publications drew much attention particularly from those involved in child protection because of there conclusions. Punt et al10 published a lengthy rebuttal the main thrust of the which suggested intrinsic flaws within the research and a lack of evidence to support the unified hypothesis.

Geddes et al11 recently replied to Punt seeking to clarify their hypothesis explaining that whilst some had severe corticospinal pathology a few had strikingly little axonal damage “...In other words, in terms of numbers of axons injured, such an injury was trivial and totally survivable. What was not trivial was the child’s response to that injury...we do not know the minimum force needed to stretch the neuroaxis...” . Yet again they extrapolate this time from the fact that only a few nerve fibres may need to be damaged means only minimal force may be required to cause this small amount of damage but that the response was catastrophic, and that the damage to a small number of axons would be in itself survivable. This sounds plausible and intuitive, but just because only a few nerve fibres are damaged (and in some cases no obvious axonal damage in this area was found) does not prove that it does not require much force to cause this damage. It would seem that it is not that in terms of the number of axons damaged that determines that it is a trivial injury and survivable but it is the location of that damage.

The main scientific finding of Geddes et al in the first two papers was that in cases of retinal haemorrhages with thin film subdurs and in the absence of other injuries that the pathological finding is more commonly that of hypoxic ischaemic encephalopathy rather than diffuse axonal injury. This seems to have been lost in the subsequent arguments over the forces required to produce these findings.

Conclusion
The cause of retinal haemorrhages, including the biomechanics of vitreo-retinal traction, raised intracranial pressure, changes in vascular permeability all remain unproven hypothesis as does the suggested mechanism of haemorrhage from Geddes. We are trying to make informed decisions on the basis of necessarily incomplete observational data, using inadequate mathematical, anthropomorphic and animal models which do not reflect the true nature of the normal infant nor the forces involved in shaking. The minimum forces required to cause such haemorrhage are not known and given that previous assumptions have been based on calculations relating to the generation of diffuse axonal injury, the use of the finding of retinal haemorrhages in isolation as a surrogate measure for the forces involved becomes dubious.

Regardless of the recent debate the observational evidence to date remains that children with non accidental injury may have no visible retinal haemorrhages, whilst non accidental injury and birth are the only circumstances in which multiple retinal haemorrhages in differing layers of the retina have been accurately documented.

This small albeit difficult group should not divert us from a willingness to evaluate the literature critically, participate in reasoned debate and in further research and certainly not detract from the main message...DON’T SHAKE THE BABY

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Note: This update represents the views of the author, who is not a member of The Royal College of Ophthalmologists Working Party on Child Abuse, and as such does not represent the views of the Royal College of Ophthalmologists.

References: