Recognition and management of Shaken Baby Syndrome

Abusive head trauma (previously referred to as Shaken Baby Syndrome) consists of a triad of findings which included subdural haemorrhage, retinal haemorrhages and encephalopathy after receiving a shake injury or blunt trauma to the head. Debate rages regarding the exact mechanism. Previously published reports on abusive head trauma (AHT) highlight the young age of the victims (median 4 months of age), the significant preponderance of male infants (3:1 in most series), the high rate of probable male perpetrators (just over 50%), and relatively high rates of mortality and morbidity 1,2,3,4,5,6,11, 12, 21.

Abusive head trauma is a leading cause of death and disability in young children, and while many descriptive studies of AHT have been published, many questions remain regarding the timing and mechanism of injury1.

The recent General Medical Council Inquiry in the United Kingdom against Dr Waney Squier (a neuropathologist from Oxford), led to a very significant media storm in both the national press and television questioning whether AHT was in fact a real entity.

The existence of AHT in infants and children is a settled scientific fact. The scientific support for AHT comes from over 40 years of research in a broad array of clinical and basic science disciplines. When an infant is subjected to rapid acceleration, deceleration and rotational forces, with or without impact, it results in a unique constellation of intracranial, intraocular and cervical spine injuries.

The annual incidence of AHT is reported at 24.6 per 100,000 infants in Scotland to 28.7 per 100,000 in Estonia 3,6. In the United States it is estimated that AHT is responsible for 80 deaths and 1,200 seriously injured infants per year. Estimated numbers of cases may be just the ‘tip of the iceberg’, as many infants with less severe forms of injury may not be identified or brought to medical attention. We have no accurate occurrence rates as yet in Ireland, but anecdotal data suggests up to one case per month.

Manifestations can be mild, moderate or life-threatening, and in mild cases, the diagnosis may easily be missed. Infants may be re-injured after missed diagnosis. The outcome is often devastating, with 15-27% of infants dying as a result of their injury and more than a third having serious neurological sequelae. Whether shaking alone can cause the severe intracranial injury associated with AHT is a topic of considerable debate, and a recent study2 concluded that blunt trauma was necessary to cause the intracranial injuries seen. Subdural haemorrhage is the commonest injury, with hypoxic ischaemic injury
being a key feature influencing the future prognosis. Spinal injury can be missed and there is frequently a lack of external evidence of trauma. Subdural haemorrhage is more likely to be bilateral but can be unilateral. Retinal haemorrhages are usually bilateral and are said to be flame-shaped, multi-layered and diffuse. Rib and metaphyseal fractures and spinal may also occur. Recent research has shown that whiplash shaking alone can lead to vitreo-retinal traction leading to retinal haemorrhage and rupture of the bridging veins which leads to subdural haemorrhage.  

In general terms, children do not behave normally immediately after AHT and show an immediate onset of symptoms. The timing of less severe AHT is much more problematic but apnoea often identifies the time of injury. The most commonly reported symptoms in AHT are limpness, seizures, vomiting, lethargy and apnoea. Therefore, if a previously well infant presents with history of a sudden decrease in consciousness or seizures and no history of injury, AHT needs to be considered in the differential diagnosis. An accurate, detailed and comprehensive history is essential. While retinal haemorrhages are not pathognomonic for AHT, they are overwhelmingly more common in AHT than in non-inflicted injury. Retinal haemorrhages may be unilateral. Those retinal haemorrhages with a high specificity for abuse include those involving multiple retinal layers, if extending out to the edges of the retina on indirect ophthalmoscopy and if signs of traumatic retinosclerosis are seen.  

A full assessment of suspected AHT includes: a detailed physical examination, a formal ophthalmological assessment (by a paediatric ophthalmologist), neuroimaging (ideally MRI) and imaging of the spine, a full skeletal survey (plus a three dimensional rib CT or repeat Chest X-ray after 10 days), and a detailed coagulation profile.  

Acute subdural haemorrhage indicates recent injury, mixed subdural haemorrhage may indicate prior abuse and chronic subdurs may indicate that the process has been evolving over a one to three week period. Interhemispheric, convexity and multiple subdural haemorrhages all have a strong association with AHT.  

There appears to be little seasonal variation except that AHT tends to cluster in the latter part of the year from October to December. Differential diagnoses include accidental falls, birth trauma, coagulopathies including haemorrhagic disease of the newborn, arteriovenous malformations of the brain, metabolic disorders including glutaric aciduria type 1, Menke’s disease, Ehler Danlos syndrome and benign extra-axial fluid collection of infancy with associated macrocephaly.  

The diagnosis of AHT should be considered if there is a combination of intracranial bleeding, retinal haemorrhage and long bone or rib fractures, macrocephaly/developmental delay, inconsistent history, or an acute life threatening event.  

In a study by Hettler et al, subdural hematomas were found in 88% of infants with AHT. In another
study, 85% children had subdural hematoma when the injuries were associated with impact, 91% with subdural hematomas with shaking only. However all children in this study had subdural hematomas when both shaken and had impact injuries\(^1\).

Retinal haemorrhages occur in 70% of children with AHT from impact injuries, 84% from shaking only and 94% with those suffering impact and shake injuries\(^1\). When assessing children for AHT, it is important to detail types of retinal haemorrhage. These may include preretinal, intraretinal and subretinal haemorrhages. It is also important to document the number, its distribution (confined to posterior pole of the retina or spreading to the ora of retina), and pattern of haemorrhages. Two-thirds of AHT victims had numerous, multi-layered retinal haemorrhages extending to the ora. Fifteen percent have no retinal haemorrhages. Absence of retinal haemorrhage does not rule out child abuse. Traumatic retinoschisis is a particularly diagnostic lesion caused by traction applied to the retina by the vitreous jelly as the child is submitted to repetitive acceleration-deceleration forces. The retina splits, creating a blood filled cystic cavity, not reported in otherwise well children except AHT victims and perhaps severe head crush injury which would otherwise be obvious by history\(^8\). Retinal haemorrhages are associated with birth mainly through the vaginal route.

These should have resolved within a month of birth\(^9\). Likewise, retinal haemorrhage occurs rarely in accidental head injury and is associated with extraordinary force\(^10\). Starling et al\(^1\) showed that skull fractures accounted for 40% of children with impact injuries, 6% of those with alleged shaking only injuries. Eighteen percent of children had skull fractures with both a shaking and impact type injury. Also, up to 27% of skeletal injuries secondary to abuse are rib fractures. Most of these are located at the posterior aspect of the rib. There is usually no signs of bruising over these areas. Other areas like the anterior and lateral aspects of the ribs suffer fractures due to direct trauma to the chest wall. Many of these rib fractures may go unnoticed if there is no displacement so it is now recommended that a repeat x-ray is carried out 10-14 days later\(^11\).

Management of AHT centres around treatment of the acute medical condition. This is complemented with a multidisciplinary team approach involving social workers both within the hospital and in the community.

Social workers should be part of the team that is initially involved with the assessment of the infant on arrival to the hospital. This prevents duplication of the interview with the parents. They also provide counselling and support for families involved with AHT. In addition, they will be key in arranging home care services, follow up care with the community agencies, short-term care placements and liaison with the external agencies to reintegrate families if appropriate.

Most experts believe that AHT is largely preventable. The most common preceding incident is sustained crying. Exhausted parents and other caregivers may become frustrated and angry when infants in their
Care cry inconsolably. Prevention strategies include identifying high risk families, increased community recognition, providing support for parents. As healthcare professionals we need to recognise exhaustion and frustration, arrange admission to hospital to defuse the situation if necessary and provide parenting education and link groups. In the words of Dr Carol Jenny, an internationally renowned expert in the field of AHT: ‘Abusive Head Trauma begins with intense frustration and anger. In its aftermath lives relationships, families and futures can be changed forever’.

Prevention is far better than cure

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