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Patterns of presentation of the shaken baby syndrome

Four types of inflicted brain injury predominate

Editor—One of the controversies that has recently arisen in cases of alleged shaken baby syndrome concerns the disparity between certain neuropathological findings at necropsy and whether these findings are consistent with the entity regarded as the shaken baby syndrome.

A database was collected for more than five years of documented Scottish cases of suspected non-accidental head injury diagnosed after a multiagency assessment and including cases with uncorroborated confessions of perpetrators and criminal convictions. Several patterns of presentation allow delineation of cases into four predominant types.

Hyperacute encephalopathy (cervicomedullary syndrome)
This hyperacute encephalopathy (6% of all cases) results from extreme “whiplashing” forces, the infant suffering the equivalent of a broken neck or, more correctly, a broken brain stem. In infants with a median survival of one day Geddes et al described localised axonal damage at the cranio cervical junction, in the corticospinal tracts, and in the cervical cord roots, consistent with hyperflexion and hyperextension movements.1 These cases, which truly reflect a “whiplash” shaking injury to the stem, are infrequently seen by clinicians because the patients are either dead on admission or die shortly thereafter.

Presentation is at 2-3 months of age, with acute respiratory failure (direct medullary trauma) and cerebral oedema (a “black brain” on imaging). At necropsy these infants have severe brain swelling and hypoxic injury but little axonal shearing and only a thin (trivial) subdural haemorrhage. Such presentations could result from a primary injury to the brain stem, induced by hyperflexion and hyperextension, or, rarely, from traumatic thrombosis of the vertebral arteries in the foramina of the cervical vertebrae.

Acute encephalopathy
An acute encephalopathic presentation (53% of cases) is characterised by a depressed conscious state, raised intracranial pressure, fits, apnoea, hypotonia or decerebration, anaemia, shock, bilateral subdural haematomas, and widespread haemorrhagic retinopathy. Coexistent rib fractures, metaphyseal fractures, or other non-accidental injuries may be found. This is the commonest presentation seen by paediatricians and is referred to as the classic shaken baby syndrome (repetitive rotational injury). Depending on whether additional signs of impact are noted (focal subdural, extradural, or subgaleal haemorrhage; scalp injury; or skull fracture), the syndrome has been referred to as the shaken impact syndrome.

The brain injury is well documented from studies of magnetic resonance imaging,2 which show widespread vascular shearing with convexity subdural haemorrhages enlarging over the first week (as well as interhemispheric, subtemporal, suboccipital, and posterior fossa subdural haemorrhages), torn bridging veins, cerebral oedema, haemorrhagic contusions and lacerations, and white matter shearing, with tears and petechial haemorrhages at the junction between grey and white matter and in the corpus callosum. Up to 60% of cases have serious long term morbidity.

Subacute non-encephalopathic presentation
In infants with a non-encephalopathic subacute presentation (19% of cases) the brain injury is less intense, without swelling, diffuse cerebral hypodensities, or clinical encephalopathic features. These children have various combinations of subdural and retinal haemorrhages, rib fractures and other skeletal fractures, bruising, etc. The outcome in this group is better.

Chronic extracerebral presentation
A chronic extracerebral presentation (22% of cases) is seen in children of a few months of age who present with an isolated subdural haemorrhage, which is often chronic (> 3 weeks) and late in presenting. A rapidly expanding head circumference and signs of raised intracranial tension are common: the child may be irritable, vomiting, failing to thrive, hypotonic, fitting but with little encephalopathy.

The primary injury is extracerebral but with potential secondary injury from raised intracranial pressure and reduced cerebral perfusion pressure and hypoperfusion, oedema, and metabolism to flow mismatch in the white matter.3 Any retinal haemorrhages originally present have disappeared by presentation. The injury has occurred weeks earlier, and its force has been insufficient to rupture the weakest bridging vein(s) but insufficient to produce an acute encephalopathy. The prognosis is good with recognition and appropriate treatment.

Clinicians will have difficulty in attributing a causative mechanism and timing to such late presenting (idiopathic) subdural haemorrhages. Only in the presence of residual features of physical abuse (such as fractures), along with identifiable risk factors, would non-accidental injury be considered. Most cases remain aetologically unexplained, although trauma remains the likely cause, but they are unlikely to be legally pursued beyond medical investigations and social work inquiry.

Conclusions
We postulate that a spectrum of clinical features is related to the intensity and type of injury in babies with inflicted brain injury, reconciling the clinical and neuropathological findings. Infants can be traumatically injured in many ways, and many instances are uninvestigated. Thus the generic term non-accidental head injury or inflicted traumatic brain injury should be used in preference to shaken baby syndrome, which implies a specific mechanism of injury.

After the history, examination, and investigations have been considered the following conclusions about the cause of brain injury can be reached: It is characteristic of, consistent with, possibly due to, or not the result of, non-accidental trauma.

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Competing interests: None declared.

Subdural and retinal haemorrhages are not necessarily signs of abuse

Editor—The "serious data gaps, flaws of logic, and inconsistency of case definition shown up by the evidence based case report of the shaken baby syndrome (p 754) and highlighted in the accompanying editorials (pp 719 and 720) will be of interest to the many parents who over the past 10 years have maintained that they have been wrongly accused and convicted of causing their children's injuries.1,2

Furthermore, the recent evidence emphasised by Geddes and Plunkett that trivial falls and other minor injuries can give rise to the allegedly characteristic signs of subdural and retinal haemorrhages is consistent with a trial of possible alternative explanations for shaken baby syndrome. This triad has emerged from an analysis of 98 parental accounts reported to the support group the Five Percenters, each of the three being compatible with a distinct type of neuropathology.

The first is minor trauma (37% of cases). This group gives a history of minor trauma (such as a fall from a bed or sofa) with either immediate loss of consciousness or delayed presentation of an acute subdural bleed and retinal haemorrhages. This is in line with the recently reported series from the United States of independently witnessed minor falls resulting in an acute intracranial bleed, the retinal haemorrhages being caused by a sudden rise in retinal venous pressure as in Terson’s syndrome.3

The second is birth injury (29% of cases). The clinical presentation in the second group is quite different. There is a general period of variable length of non-specific symptoms such as vomiting and lethargy warranting repeated medical consultations until computed tomography shows the presence of a chronic subdural haemorrhage. The most likely aetiology is a subdural bleed at birth, which, though usually associated with prematurity or a difficult delivery, can follow a normal delivery.4

The third is respiratory arrest (22% of cases). In this group the precipitating event is suggestive of respiratory arrest—often followed by attempts at resuscitation—that could result in the subdural and retinal haemorrhages characteristic of hypoxic encephalopathy. The findings that severe traumatic brain damage is not, as previously thought, present in these cases contradicts the assumption that such injuries could only have been induced by violent shaking.4

A fourth type of presentation, epileptiform seizures (12%) is presumably secondary to underlyng intracranial disease—and is thus uninformative about possible aetiology. These three patterns of clinical events—in the absence of other circumstantial evidence for non-accidental injury—offer a more credible explanation than shaken baby syndrome for the presence of subdural and retinal haemorrhages. It should be noted that shaking has never been directly observed or proved to cause such injuries but is rather an inference based on (contested) theories of biomechanics.5 By contrast, consistent parenteral testimony tallies with descriptions from independent witnesses. Furthermore, each pattern of clinical events is consistent with a distinctive type of neuropathology of acute subdural, chronic subdural, or the thin subdural hypoxic encephalopathy.

While we recognise the limitations of the volunteered parental testimony on which this analysis is based, the same triad of presentations—designated as acute encephalopathic, idiopathic subdural, and hyperacute presentation—has also been independently identified from an extended database of cases of suspected non-accidental injury (see previous letter).6 These findings necessarily raise disturbing questions about the validity of the opinions expressed by medical experts in the courts. They warrant further, urgent, and appropriate scientific investigation.

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Competing interests: None declared.


8 Minna KA, Burwell A. Patterns of presentation of shaken baby syndrome. Electronic response to: Brain haemorrhage in babies may not indicate violent abuse. bmj.com 2003; bmj.com/cgi/directx/323/7590/6706a (accessed 17 July 2005).

This general reluctance must be for several reasons

Editor—In his news item Dyer reports that doctors are reluctant to work on child protection committees.1 I have yet to meet a paediatrician who is genuinely keen to do child protection work. Not surprisingly, the Royal College of Paediatrics and Child Health is experiencing enormous difficulties filling the relevant posts.

Most paediatricians in training today do not wish to do community paediatrics in the future. It is certainly essential to have a named paediatrician for child protection in every hospital, but, ironically, in my experience, even the named paediatricians for child protection in some cases are reluctant to show passion in this field.

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measures was there a significant difference in medical intervention overall. One conclusion not emphasised is that labour in the pool is associated with significantly more neonatal morbidity, with six babies from this group admitted to special care and none from the standard augmentation group (P = 0.013).

Inadequate numbers may be responsible for the absence of a significant difference in epidural rates. As discussed by Cluett et al, recruitment to randomised controlled trials of obstetric intervention is often difficult. Many women have preconceived ideas about how they would like their labour to be managed and are unwilling to be randomly allocated management.

It is therefore particularly unfortunate that the authors’ attempts to address this important question seem not to have been supported by local policy makers. The unit’s adoption of a more conservative approach to augmentation half way through the study seems to have been based on pre-existing research rather than any contemporaneously published report. Surely it would have been ethical to delay such a policy change until the researchers had completed recruitment?

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Competing interests: None declared.

The editor confesses—and apologises to anybody upset
I wrote the words on the cover of the BMJ of 7 February.

Initially I wrote: “Labouring in water.” Unfortunately it made only one line, and we needed two. I thus had to change it, and my first thought was “Giving birth in water.” But, I wondered, did these women give birth in water?

I quickly scanned the paper, and I didn’t find the answer.

Then I wondered if there was that much difference linguistically between “labouring” and “giving birth.” Do people think of “giving birth” as the moment of birth or the longer process?

Then something else happened—as it always does—and I left it as “Giving birth in water.”

I apologise to anybody who thinks it horribly wrong, but this little story illustrates the exigencies of putting a journal together.

Richard Smith
editor
BMJ

Competing interests: I’m the editor of the BMJ and accountable for all it contains.


Cover wrongly said study was about giving birth in water
Editor—The study by Cluett et al reinforces anecdotal experience that allowing a woman in labour to have a good soak in a deep bath will relieve pain and facilitate labour.1 Indeed, it is a shame that the traditional big bath has been removed from the labour ward.

However, readers will have been misled by the headline on the cover of the BMJ—"Giving birth in water.” The study is about labouring in water, not delivery. I don’t know how many of the women chose to deliver in the water bath, but from my quick scan through the paper I do not think that this was the intention. I note that there was a mean delay of six hours (range 2-10 hours) between women leaving the pool and giving birth.

This study is on water immersion as an option for women in the first stage of labour, not about delivery in water.

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Competing interests: None declared.


Folic acid as ultimate in disease prevention
Editor—Lucock considered the likely effects of mass use of folate but did not mention the potential benefits to mental health.1 Associations between folate status and mood have been known for some time, with folate deficiency considered a treatable cause of depression. Emerging evidence from randomised trials shows that the augmentation of conventional antidepressant treatments with folate may improve outcome, and this effect may be seen even in patients with normal folate concentrations at baseline.2 Observational studies also find associations between folate status and dementia,3 although currently trials of dietary supplementation are not conclusive.4

The effects on health of fortification with folate may not be limited to birth defects, vascular disease, and cancers.5

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Competing interests: None declared.

Folate also improves mental health
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Competing interests: None declared.
Beware of vitamin B12 deficiency

Editor—In response to the review by Lucock on folic acid supplementation,1 we have a word of caution about possible concomitant vitamin B12 deficiency, which may also cause raised concentrations of homocysteine.

Vitamin B12 deficiency can be subtle, manifesting only as an increase in concentrations of homocysteine and methylmalonic acid in blood and urine, with concentrations of vitamin B12 at the lower limit of normal.2 Vitamin B12 concentration varies in different populations. The US NHANES III survey found a mean serum B12 value of 318 pg/ml, and 3% of the population had a concentration of less than 200 pg/ml.3 In Israel we reported a vitamin B12 deficiency of 30% in 130 serial patients undergoing coronary angiography.4 Health maintenance organisations in Israel responded to the widespread deficiency of vitamin B12 by lowering the normal values of their laboratories.

Since folic acid supplementation may be harmful in the presence of undiagnosed vitamin B12 deficiency, we recommend that vitamin B12 concentrations be determined before administration of folic acid. Another approach may be to use multivitamin tablets. The use of a “folate” supplement consisting of 1 mg folic acid, 400 μg vitamin B12, and 10 mg pyridoxine both reduces the concentrations of homocysteine and decreases the rate of restenosis after angioplasty.5 The cost effectiveness of these approaches may differ from country to country, depending on the prevalence of vitamin B12 deficiency.

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Competing interests: None declared.

1 Lucock M. Is folic acid the ultimate functional food?—a rationale for disease prevention?BMJ 2004;328:211-4. (21 January)

Websites on screening for breast cancer

Language may be as misleading as statistics

Editor—Ironically, the paper by Jørgensen and Gøtzsche purporting to reveal “bias” in government and advocacy websites provid-
Eysenbach yawns at our study at our exit, as well as the principle of informed consent by omitting information on the major harms of screening. This is hardly something to yawn about but calls for a change.

We thank Hanson for drawing our attention to our error, for which we apologise. Both of us independently and correctly classified the National Women’s Health Network as an organisation that does not accept industry funding, but a mistake occurred during data transfer for statistical analysis. Since Hanson notes that the organisation is not only an advocacy group but also a consumer group, it seems most appropriate to exclude it from the analyses that compare consumer groups with other groups. This correction would lead to the same or very similar P values as those we reported in our paper, and our conclusions remain unchanged.

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Competing interests: None declared.


GPs need training and funding in caring for refugees and asylum seekers

Enr0r—UK asylum seekers and refugees have the same rights to health care as the settled population. One less, the refugees report difficulties accessing health care—for example, in registering with a general practitioner.1 They are also sometimes registered as temporary residents, which is detrimental for continuity of care.2

We sent a postal questionnaire to all 129 general practitioners in Lothian, an area in Scotland with an estimated 3000 refugees. It focused on the need for extra funding for general practices with refugees, the best place for providing primary care services, and the need for training.

Ninety-five responses were received (a response rate of 73.6%). The table summarises the results (see bmj.com for more details).3

<table>
<thead>
<tr>
<th>Question</th>
<th>No (%) giving answer</th>
</tr>
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<tbody>
<tr>
<td>Do you have any experience of treating asylum seekers or refugees?</td>
<td>Yes 94 (93)</td>
</tr>
<tr>
<td>Have you undergone special training to care for asylum seekers and refugees?</td>
<td>Yes 94 (93)</td>
</tr>
<tr>
<td>Does your practice staff undergo any training to care for asylum seekers and refugees?</td>
<td>Yes 94 (93)</td>
</tr>
<tr>
<td>Do asylum seekers and refugees have the same rights for provision of NHS treatment?</td>
<td>Different rights 93 (11)</td>
</tr>
<tr>
<td>Do asylum seekers and refugees have to pay for NHS treatment?</td>
<td>Yes 94 (93)</td>
</tr>
<tr>
<td>Are asylum seekers and refugees registered as temporary or permanent residents?</td>
<td>As temporary residents 94 (93)</td>
</tr>
<tr>
<td>Do you require asylum seekers or refugees to produce any proof of identity for registration?</td>
<td>Yes 86 (86)</td>
</tr>
<tr>
<td>Where do you feel that asylum seekers and refugees should be given primary care services?</td>
<td>GP 86 (86)</td>
</tr>
<tr>
<td>Are there any general practitioners supporting extra funding if they have a high caseload of asylum seekers and refugees?</td>
<td>Yes 94 (93)</td>
</tr>
</tbody>
</table>

Many non-respondents cited lack of experience with refugees as a reason for not answering.

*General practitioner.

1 Many non-respondents cited lack of experience with refugees as a reason for not answering.

About one third of general practitioners (86%) who had not received training, 17 (21%) wanted training. Nearly one fifth were unsure or incorrect about refugees’ entitlement to free NHS treatment. Respondents were divided on whether refugees should be treated at normal practices or by specialist services. Most general practitioners thought that practices with a high caseload of refugees should receive additional funding. Thirty one general practitioners’ suggestions on funding per refugee ranged from £20-£1000, with a median of £151 and a median of £100. General practitioners supported extra funding and suggested about £100 per refugee; many favoured treating refugees in normal practices, but many had no relevant training. Some were unaware of refugees’ NHS entitlements, as has been previously reported.4 To our knowledge, these are the first published data on the views of general practitioners (or their international equivalents) on funding, training, and the place of care for refugee primary healthcare. A need exists to develop approaches to health care in urban centres with varying numbers of refugees.

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Competing interests: None declared.

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4 Dar S. General practitioners’ knowledge of issues relating to asylum seekers is poor. BMJ 2000;321:893.

Medicine’s weapons of mass destruction come in human form

Editor—In his Soundings article on weapons of mass destruction Loeffler uses few words to say much.5 He says that now the biological enemy is microscopic and sub-microscopic. However, when talking of weapons of mass destruction, we should give credit where credit is due.

Our real enemies are the powers that aim to make us dependent: the merchants who try to sell health care as a proactive entity rather than a reactive one. They offer free examinations, and they manipulate long established laboratory measurements, all in the name of more profit. They go hunting for potential new patients and persuade them that they need treatment or "prescriptive" measures. They deliver “health care” to the eager, brainwashed consumer like the milkman delivers milk. But they often come empty handed. They do not owe you anything, but they promise much.

Weapons of mass destruction are hard to find in Iraq; modern medicine they are abundant (if cosmetics enhanced).

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Competing interests: None declared.