## SHAKEN BABY SYNDROME (SBS) AND NON-ACCIDENTAL INJURIES (NAI) ----DR MARK DONOHOE

This document is a de-identified extract from medical reports prepared in 1999 by Dr Mark Donohoe in selected and typical cases of alleged shaking and injuries to infants. It's purpose is to provide a framework for assessing assumptions and evidence required for determining that particular injuries were caused by intentional and violent shaking. It is accompanied by a more general assessment of the "quality of evidence" in the research so far published (to late 1998) in the peer-reviewed medical literature, along with a listing of the references reviewed for both SBS/NAI and Terson's syndrome.

## **1 THE "SHAKEN BABY SYNDROME"**

It is not possible to address the specifics of any particular case without addressing the general medical and scientific literature regarding the so-called "shaken baby syndrome" (SBS). I have performed such a review of the medical literature in order to assess the quality of evidence related to the diagnosis of SBS, and include this as Appendix I, along with the references reviewed. The question which needs to be answered is not, "could this be a case of shaken baby syndrome?", because this is clearly one possibility. The questions which need to be answered would appear to be:

1/ "could this case represent anything other than SBS?"; and the related question

2/ "how certain are we that this was a case of SBS, as opposed to anything else?" This report seeks to reach an objective answer to these questions in this particular case.

## **1.1 HISTORICAL ISSUES AND EVIDENCE FOR SBS**

For doctors dealing with potentially abused children, the error of assuming abuse, even when none has occurred, is acceptable and (some would argue) necessary. It is certainly preferable to the alternative of failing to identify abuse which has occurred. In the past, the medical profession seems to have failed to identify cases of abuse, resulting in tragic outcomes for families and for the children not identified in particular. In the last decade or so, a concerted effort has been made to address this historical blind-spot, and a number of advocacy groups for the rights of the child have emerged. Public educational campaigns have arisen, bringing the risks of shaking to the attention of the public, and the medical profession, particularly in hospital emergency settings, tends to scrutinise all cases of childhood injury or unexplained illness for evidence of abuse. On the whole, this has been a positive change, and may have resulted in decreases in death or injury of babies and children, although this has yet to be demonstrated.

As with all "swings of a pendulum", however, there are associated dangers of such increased surveillance for abuse. The most obvious risk is that cases not associated with abuse will be incorrectly assumed to be abuse, and that the totality of evidence in cases will be ignored in favour of apparently "incriminating" evidence. I say this because it is, even in theory, impossible to attain perfection in the correct attribution of cause of injury. In the past, actual abuse cases were missed as a result. Now, cases unrelated to abuse are occasionally incorrectly attributed to abuse. The medical tests which are supportive of the diagnosis of non-accidental injury (NAI) have been pushed by many advocates as being proof of NAI generally, and SBS in particular. The diagnosis of NAI and SBS rests on a number of factors, including medical and social history, family circumstances, medical examination, a wide range of test results, and the results of investigations and interviews by experienced multi-disciplinary teams. The diagnosis of NAI or SBS cannot ever rest upon the results of a few isolated investigations alone, as has been the case in recent years.

There are two main reasons for pursuing suspected NAI vigorously, namely: identifying and punishing any perpetrator, should the person be found; and decreasing further risk to the abused child and other children in the family. While the medical profession may well be acting in the interests of the child by assuming NAI even when it has not occurred, one must recognise the potential for harm to the family and child where such assumptions prove to be incorrect. The decision on whether or not NAI has occurred, and who is responsible for it if it has occurred, must be made by the legal process, and such judgements must be dispassionate and based on evidence, rather than assertion by those wishing to protect the child. While it is self-evident that the safety and rights of the child must be protected in every case, the rights of the parents and carers cannot be ignored or forgotten in so doing. There is a clear risk in assuming that abuse has occurred in cases where the evidence is weak, and where alternative opinions have not been adequately pursued and excluded. The cases which I have so far reviewed would never have been investigated for potential abuse had it not been for the medical findings, as there was no evidence of or suggestion of abuse. In such cases, NAI has been assumed where other alternative explanations not only exist, but are arguably more likely than NAI.

Finally, the tendency of the medical profession to ignore the possibility of adverse reactions to vaccines, especially those containing thiomersal (a mercury preservative), is important. In more than half of the cases I have reviewed, there is an apparent temporal relationship between the injury and prior vaccination with thiomersal-containing polyvalent vaccines or shaken baby syndrome (SBS) & non-accidental injuries (NAI) review . Dr Mark Donohoe . 20/8/01 Page 2

multiple simultaneous vaccinations. The vaccine typically leads to pyrexia and crying immediately following vaccination, and the infant is given paracetamol (acetaminophen) at increasing doses for control of crying and pyrexia in the following days. In most of these cases, a broad spectrum antibiotic has also been prescribed in the same period. It has been suggested that the alternative explanation should be preferred, namely that the vaccination itself caused no injury, but induced crying which lasted for days, leading to frustration and eventual shaking by a parent or carer. Even if this is accepted, it raises a most interesting issue of ultimate causation. Had the vaccine(s) not been administered, the adverse reaction and crying would not have occurred, and there would have been no risk or likelihood of shaking or injury of any type. The acceptance of such crying and pyrexia as a "normal and expected" consequence of polyvalent vaccinations in up to 20% of infants does nothing to address the consequences of the crying and the increased risk of injury to the child from such a common event. It is disingenuous to argue that "the disease would have been worse than the vaccination", because the disease may or may not have occurred, and is a part of normal life of a child, whereas vaccination is a procedure requiring informed consent. If one identifiable risk of vaccination is that of shaking and injury following prolonged crying (assuming that shaking did occur as a result of the vaccine-induced crying), then this needs to be conveyed to parents of children being vaccinated as part of the informed consent. If they are made aware of the potential for prolonged crying and pyrexia, and are told of the risks associated with shaking which may be a consequence of this crying, then the risk itself may be reduced.

Thus, it is arguable that vaccination is an independent predictor (or risk factor) for neurological injuries in infants, whether it is directly causative or predisposes to other outcomes which are causative. A recognition of the association (direct or indirect) may allow for action and warnings which may decrease the likelihood of adverse outcomes.

#### **1.2 AXIOMS AND ASSUMPTIONS REGARDING ALLEGATION OF SBS**

The case for this infant being the victim of intentional shaking rests upon certain axioms and assumptions, which may be reduced to the following:

1/ subdural haemorrhage (SDH) does not occur in a normal, healthy infant;

2/ SDH combined with retinal haemorrhage (RH) is pathognomonic of "non-accidental injury" (NAI);

3/ in the absence of identifiable external trauma, SDH and RH are only caused by

violent "acceleration/deceleration" actions caused by an adult, otherwise known as "shaken baby syndrome";

4/ SBS must be intentional, or at least they would appear intentional and excessive to any dispassionate observer;

5/ the diagnosis of "shaken baby syndrome" can be made with absolute certainty on the basis of ophthalmological and radiological assessment alone. shaken baby syndrome (SBS) & non-accidental injuries (NAI) review Dr Mark Donohoe . 20/8/01 Page 3

6/ the severity of shaking required to cause these findings would have been obvious, and was unrelated to any shaking or corporal punishment admitted to by family members. I shall address each of these statements separately, and attempt to assess the evidence for and against each statement.

## **1.2.1 SPONTANEOUS SUBDURAL HAEMORRHAGE (SDH) DOES NOT OCCUR IN A NORMAL, HEALTHY INFANT**

Clearly, SDH should not occur in a "normal healthy infant", although there is no medical or scientific evidence to back this assertion one way or the other. There is a logical difficulty, of course, in that any such cases which did occur in a "normal healthy infant" would almost certainly be attributed to NAI, whether or not there was other evidence to support such an attribution. This is a part of a broader defect of logic in NAI cases, that all unexplained injuries can be assumed to be NAI, which has been allowed to continue in an effort to protect the child. There is evidence that SDH does occur in normal, healthy neonates as a result of birth trauma, and even in cases of normal, uncomplicated vaginal delivery. There is clear evidence that SDH can be caused by anatomical, infectious and biochemical disorders which cause no obvious symptoms prior to the intracranial bleeding. In such circumstances, the infant appears "normal and healthy" (although probably not robustly healthy) despite the underlying, predisposing pathology.

Such disorders, predisposing to SDH, include:

1 arteriovenous malformations (AVM) or shunts (AVS);

2 certain infections such as malaria, hepatitis, septicaemia;

3 intravascular coagulopathy due to bacterial endotoxin;

4 certain malignancies, such as leukaemia;

5 clotting disorders;

6 accidental or intentional poisoning;

7 liver disease, such as microvesicular steatosis, hepatitis, etc;

8 gastrointestinal disorders, causing malabsorption, affecting trace elements, ascorbate or fat soluble nutrients (esp vitamins E and K);

9 metabolic disorders, such as Reye's syndrome, diabetes, disorders of energy metabolism;

10 inherited disorders, such as enzyme defects or chromosomal defects;

11 vascular and connective tissue disorders.

This list is not intended to be exhaustive. It is also possible that more than one predisposing factor existed, and that it was a combination of factors which led to the intracranial haemorrhage. For example, a mild factor XIII deficiency, when combined with shaken baby syndrome (SBS) & non-accidental injuries (NAI) review . Dr Mark Donohoe . 20/8/01 Page 4

malabsorption or Reye's syndrome, would be likely to result in a greater risk of intracranial bleeding than would any one of these factors alone. In an infant under six months, such disorders may be asymptomatic, or may simply result in a non-specific failure to thrive. In some of these diseases, the first presentation is that of an intracranial bleed. It is only possible to exclude these other causes by careful history taking, examination, appropriate and extensive testing, including assessment of nutritional status, enzyme levels, and assessing pathology of the liver, gut, kidneys, muscle, brain and heart. It is my view, based on the cases I have reviewed, that there is a tendency to assume that NAI has occurred in infants with SDH and RH, and as a consequence of that assumption, there is a general failure in the first instance to pursue other potential causes or contributions to the injury as vigorously as would be expected.

Further, once the allegation of abuse has been made, and police or welfare services have become involved, there is a general unwillingness to consider any other plausible causes of the injury. In a sense, the parents or carer are assumed to be guilty, and have no logical way of "proving" their innocence. Investigations are continued, even if the infant dies, but those investigations are usually directed at building a stronger case for NAI (such as bone scan, MRI, autopsy), rather than identifying alternative causes or contributory factors.

## 1.2.2 SDH COMBINED WITH RETINAL HAEMORRHAGE (RH) IS PATHOGNOMONIC OF "NON-ACCIDENTAL INJURY" (NAI);

This is frequently the claim made by medical experts involved in the prosecution of SBS cases. Unless this relationship is proven, there is reasonable doubt that anyone shook or otherwise injured this infant. The term "pathognomonic" implies a two-way relationship between the symptoms and signs on one hand, and the disease in question

on the other hand. Pathognomonic symptoms or signs not only allow recognition of a disease, but differentiate it from all other diseases or disorders. Technically, it implies 100% specificity for the tests establishing the diagnosis. The combination of SDH and RH strongly support a diagnosis of NAI, but in no way constitute proof that NAI occurred. The value of the diagnosis of SDH and RH is that their presence can markedly increase the confidence of a diagnosis of NAI, given other circumstances or findings which may indicate NAI. Even if we assume that all non-accidental injuries involving shaking or blunt trauma to the head of babies cause subdural haemorrhage and retinal haemorrhage, it does not follow that all cases of SDH and RH are caused by NAI. shaken baby syndrome (SBS) & non-accidental injuries (NAI) review . Dr Mark Donohoe . 20/8/01 Page 5

One must know what other diseases or circumstances may cause SDH and RH. Since the mechanism of RH is unknown (Riffenburgh 1991), it is important that the specific hallmarks of NAI be delineated, as proposed recently (Rohrbach 1997). Rohrbach (1997) has stated,"Intraretinal haemorrhages alone are typical, though not pathognomonic for the 'battered-child syndrome'''.

According to Rohrbach, the combination of the following provides increased certainty of NAI, although there are clearly shortcomings in this single case study: 1 retinal haemorrhage; 2 crater-like appearance of central retina; 3 haemorrhagic retinoschisis; and 4 intrascleral haemorrhages in the area of the circle of Zinn-Haller. The ophthalmologists examining the infant needs to be questioned as to whether the changes seen in the particular case match these proposed criteria.

Conditions apart from NAI which may result in SD and RH include: bleeding disorders; meningitis; septicaemia; leukaemia; galactosaemia; hypertension; and Henoch-Schonlein purpura. RH may also occur simply as a consequence of the intracranial bleeding. A number of papers which deal with the mechanism of the RH have suggested that the increasing intracranial pressure and subarachnoid haemorrhage lead to retinal haemorrhages (Jacobi 1986), and these may be accompanied by SDH.

In fact, this association is known as "Terson's syndrome". A number of papers (Giangiacomo 1985, Weingeist 1986, Jacobi 1986,Keithahn 1993, Poepel 1994) point out the similarities between Terson's syndrome and the retinopathy of shaken baby syndrome, with some suggesting that SBS should be considered in the differential diagnosis of Terson's syndrome. Thus, there are other plausible and reasonable explanations for the combination of SDH and RH in an infant. Any of the factors listed above as potential causes of SDH must be considered a plausible cause of the RH as well. The presence of SDH and RH is insufficient to prove any particular

cause. It is likely that themajority of cases in which SDH and RH are found in infants under one year of age are NAI, but this is a statistical association. In any given case, these findings are only supportive of NAI. The case must stand or fall on other factors which would lead one to suspect NAI. shaken baby syndrome (SBS) & non-accidental injuries (NAI) review . Dr Mark Donohoe . 20/8/01 Page 6

### 1.2.3 IN THE ABSENCE OF EVIDENCE OF IDENTIFIABLE EXTERNAL TRAUMA, SDH AND RH ARE ONLY CAUSED BY VIOLENT "ACCELERATION/DECELERATION" ACTIONS CAUSED BY AN ADULT, KNOWN AS "SHAKEN BABY SYNDROME";

Most studies show a high proportion of NAIs are associated with other signs of trauma or abuse, either in the damaged infant or in other family members. In a recent British retrospective study (Jayawant 1998), about 60% showed signs of other trauma (bruising, fractures, resolving old SDH, etc). Other authors have suggested similar or higher percentages as showing signs of trauma or abuse, especially in cases of severe cerebral damage and death (Alexander 1990, Duhaime 1987, Lancon 1998). As noted above, factors which may cause or contribute to SDH or SAH in an infant are likely to cause RH as well. A recent study (Jayawant 1998) noted an 80% association between SDH and RH, suggesting that RH is not an independent risk predictor, but a marker of severity and extensiveness of intracranial bleeding.

A number of authors (Duhaime 1987, Lancon 1998, Closset 1992) have suggested that shaking without impact does not generate sufficient forces to cause the types of injuries seen in SBS cases. Many others disagree, holding that shearing forces tend to rupture the fragile veins across the dural space.

These other authors propose ways in which shaking with a rotatory component, possibly at particular frequencies, or with rapid deceleration caused by soft impact (eg pillow, etc), could cause such shearing. In fact, if this view of rotation at particular frequencies is correct, it is likely that the forces required to cause the damage observed may be less important than had previously been thought.

There is no case definition for SBS, and this tends to cause problems in deciding if a particular case is one of SBS. Many authors have taken the view that any unexplained presentation of SDH and RH in an infant is NAI until proven otherwise. While useful as a medical "rule of thumb", it should be noted that this leads to a reversal of the burden of proof if applied in a legal setting, and can result in the assumption of guilt in blameless carers. If all unexplained cases are defined as NAI or SBS, then the circular definition results in carers having to prove their innocence, which is logically impossible for this condition. The essential ingredient of SBS would appear to be

strong evidence of shaking and NAI. The majority of studies, however, assume that all unexplained cases of SDH and RH are SBS and NAI, and do not determine the degree of confidence that the assumption is correct.

In addition, the actions said to be required to cause NAI have changed over time from fore-aft shaking with impact, to severe and prolonged fore-aft shaking without impact, to rotatory acceleration-deceleration without impact, and more recently to rotatory acceleration-deceleration with soft object impact. shaken baby syndrome (SBS) & non-accidental injuries (NAI) review . Dr Mark Donohoe . 20/8/01 Page 7

Without a clear case definition, and without a means of proving whether an unobserved carer performed the particular actions required, the attribution of unexplained SDH and RH to SBS is neither provable nor disprovable. It is not a question which can be scientifically decided with current knowledge and techniques, and does not meet Popper's test of a valid scientific hypothesis (that an experiment can be defined which could disprove the hypothesis, were it to be incorrect). The diagnosis rests on the presence of sufficient "other factors" which would raise suspicion of NAI, and the clinical, ophthalmological and radiographic findings become supportive, rather than diagnostic, in such circumstances.

# **1.2.4 SBS MUST BE INTENTIONAL, OR AT LEAST THEY WOULD APPEAR INTENTIONAL TO ANY DISPASSIONATE OBSERVER;**

This is, again, neither provable nor disprovable. Any concept of intention is not a medical or scientific issue, as it can only be decided by the person accused of shaking. As such, this becomes a matter for determination through the legal process. As the issue is one of degree (some degree of shaking and head movement in infants being an aspect of normal play and day to day life), the matter cannot be decided as it could for impact injuries, suffocation or penetrating injuries (which are not part of a continuum of normal life activities). There are few data or cases in which the precise actions or forces used to create the injuries have been determined or observed. The lack of observational or experimental data make it impossible to determine what an independent, dispassionate observer would perceive. It may be a reasonable assumption that the rareness of the SBS-type injury (estimated incidence in Britain at less than one in 4,000 children per year-Jayawant 1998) would imply excessive force. This may be a false assumption, however, if the damage arises from a particular type of action, rather than simply being a function of applied force. Rhythmic rotation of the baby at a particular frequency, for example, would be expected to lead to an amplification of applied force and relative motion between brain and skull in the infant. Such shaking may appear to be benign, yet cause significant damage to the dural vessels.

## 1.2.5 THAT THE DIAGNOSIS OF "SHAKEN BABY SYNDROME" CAN BE MADE CONFIDENTLY ON THE BASIS OF CLINICAL AND X-RAY ASSESSMENT ALONE.

As noted above, typical clinical and radiological changes seen in SBS cannot "make the diagnosis", but can strongly support the diagnosis made by other means. If the social and family circumstances are suggestive of NAI, then the medical investigations and clinical findings can increase the confidence that a NAI has occurred, and can help define the likely process of the NAI. The question arises as to how one goes about excluding a diagnosis which has no formal definition, and no unique defining characteristics. shaken baby syndrome (SBS) & non-accidental injuries (NAI) review . Dr Mark Donohoe . 20/8/01 Page 8

A recent paper (Jayawant 1998) defines a number of characteristics of supposed and "proven" NAI in children in a retrospective study, and the findings of this would appear to suggest a set of criteria which, if applied, may increase the precision of the diagnosis.

Nine factors suggesting NAI are identified in children who have suffered SDH, namely: Social and Family Issues 1. Sex of the child allegedly abused? (two thirds are male) 2. Sex of the alleged abuser? (four fifths are male) 3. Is there a past known history of abuse of this child or siblings by this alleged abuser? (about one eighth have previously abused)

4. Is the explanation/history internally consistent? (over half of carers change their stories two or three times)

5. Did the alleged perpetrator admit to shaking? (about half do, eventually) Investigations and clinical findings

6. Haemoglobin at presentation less than 10 g/L (seen in half of NAI cases)

7. Skeletal survey (positive in 60% of NAI cases tested)

8. Evidence of some trauma or previous trauma (seen in about 60% of NAI)

9. Retinal haemorrhages (present in 80% of cases) Of the nine proposed key factors in identification of NAI, the number found in any particular case may be important in determining likelihood of SBS/NAI.

#### 1.2.6 THE SEVERITY OF SHAKING REQUIRED TO CAUSE THESE FINDINGS WOULD HAVE BEEN OBVIOUS, AND WAS UNRELATED TO ANY SHAKING OR CORPORAL PUNISHMENT ADMITTED TO BY FAMILY MEMBERS.

This can only be decided from an assessment of the social and family circumstances, and a knowledge of the family's past history and events around the time of the alleged injury. Evidence would be required that one of those with access to the infant in the period during which the injury is thought to have occurred had previously shaken or abused either the baby or another member of the family.

Additionally, the person would need to be proven to have the strength necessary to shake the child in the manner thought to be required to induce the injury. Without such evidence, and based solely upon the presence of subdural haemorrhage and retinal haemorrhage, it would seem that the attribution of the pathology to intentional abuse cannot be sustained. shaken baby syndrome (SBS) & non-accidental injuries (NAI) review . Dr Mark Donohoe . 20/8/01 Page 9.

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[Home] [Shaken Baby Syndrome]