External Hydrocephaus: A Probable Cause for Subdural Hematoma in Infancy

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Subdural hemorrhage is common in infancy, particularly in the first year of life. The most common cause is nonaccidental (child abuse), with accidental in second place.

We present three healthy infants, ages 4, 5, and 7 months that, during an evaluation for macrocephaly, were found to have frontal subdural hematoma in association with prominent extracerebral cerebrospinal fluid spaces (external hydrocephalus). There was no history of trauma or risk factors for child abuse. Skull surveys and ophthalmologic examinations were normal. All infants were neurologically intact and achieved normal developmental milestones in one-year follow-up.

We suggest that some infants with external hydrocephalus may be at risk for development of subdural hematoma with minimal or no trauma, most likely secondary to stretching of the bridging veins in the unusually widened subarachnoid spaces. Child abuse, although it should always be kept in mind and should be excluded, may not be the most common cause in this specific context. © 2003 by Elsevier Inc. All rights reserved.


Introduction

Subdural hemorrhage is common in infancy, particularly in the first year of life, and presents a major diagnostic challenge. Most cases are due to child abuse, and only in a few cases is the cause unknown [1,2].

We present three healthy infants ages 4, 5, and 7 months old that, during an evaluation for increased head circumference, were found to have frontal subdural hematoma in association with prominent extracerebral cerebrospinal fluid (CSF) spaces (external hydrocephalus). In the absence of any evidence of previous trauma, coagulopathy, or child abuse, we suggest that the enlarged subarachnoid spaces in those infants may have been a predisposing factor for the developing of subdural hematoma.

Case Reports

Case 1

A 4-month-old infant was referred to our clinic for evaluation of increased head circumference. Pregnancy, labor, and delivery were normal. Head circumference at birth was 35.5 centimeters, which is at the 70th percentile. There was no history of postnatal infection or trauma, and no factor that suggested abuse. The physical examination demonstrated weight and height at the 50th percentile. The head circumference was 45 centimeters, which is at the 95th percentile. The anterior fontanel was soft and flat. The infant was alert and was responsive appropriately to visual and auditory stimuli. Eye examination including dilluted fundus examination was normal with no evidence of papilledema or retinal hemorrhage. Neurologic examination was normal, and developmental milestones were appropriate for age. Brain magnetic resonance imaging (MRI) demonstrated bilateral prominent extracerebral CSF spaces with moderate ventriculomegaly, and small right frontal subdural hematoma (Fig 1). Laboratory studies including hemogram, serum electrolyte, liver function tests, and coagulation studies were all normal. Skeletal surveys were normal. A follow-up MRI at 1 year of age showed disappearance of the hematoma. The infant remained neurologically intact and achieved normal developmental milestones in one-year follow-up.

Case 2

A 5-month-old healthy boy was evaluated for increased head circumference, which was first noticed at age 3 months. He was born after an uncomplicated pregnancy and had been developing appropriately. There was no history of any recent trauma, vomiting, change in behavior, or lethargy. Physical examination revealed weight and height at the 75th percentile, with head circumference of 46 cm, which is at the 95th percentile. The infant was otherwise alert and playful. Complete eye examination was normal. He had a normal tone with no asymmetry, and his development was intact. Brain MRI revealed prominent subarachnoid...
spaces over both frontal lobes with 1 cm subdural hematoma over the left frontal convexity (Figure 2). Laboratory studies including hemogram, serum electrolyte, liver function tests, and coagulation studies were all normal. Skull surveys were normal. The infant continued to develop appropriately. A follow-up MRI done 6 months later was normal.

Case 3

A 5-month-old healthy boy was evaluated for increase in his head circumference. Pregnancy and delivery were normal without complications. Head circumference at the age of 2 months was at the 95th percentile, and a head computed tomography scan (CT) done at the same age indicated only prominent extra-axial fluid spaces. In the 2 months before his visit, there was a further increase in his head circumference. There was no history of head trauma, and he was otherwise asymptomatic. Physical examination revealed weight and height at the 25th percentile, with head circumference of 46.5 cm, which is just above the 95th percentile. The head was asymmetric with flattening of the left parieto-occipital region. The infant was alert and maintained good eye contact. Funduscopic examination was normal with no evidence of papilledema or retinal hemorrhage. Neurologic examination was normal and his development was appropriate. A second head CT, including three-dimensional imaging, followed by a brain MRI revealed benign enlargement of the subarachnoid spaces, with right frontoparietal subdural hematoma. There was flattening of the left parieto-occipital region but with no evidence of craniostenosis (Fig 3). Hemogram, serum electrolyte, liver function tests, and coagulation studies were all normal. Skeletal surveys were normal. The subdural collection was surgically drained. The infant remained intact neurologically and continues to achieve normal developmental milestones.

Discussion

Many studies in the past illustrate that the majority of cases with subdural hemorrhages in children under 2 years of age are due to child abuse [1,2].

The clinical presentation in those infants is nonspecific [3]; the prognosis is poor with high percentage of deaths and long-term neurologic sequel [2,4,5].

In a study of 84 infants under 1 year of age with head injury, Billmire found child abuse as a cause in 18 (95%)}
out of 19 children with intracranial hemorrhage [6]. Duhaime, in a study of 100 children under 2 years of age with head injury, found 16 infants with subdural hematoma, 13 (81%) of which were due to inflicted injuries [7]. Jayaward et al. performed a population-based study of children under the age of 2 years who presented with subdural hemorrhage. Twenty-seven (82%) out of 33 cases were highly suggestive of abuse [2].

The finding of subdural hematoma resulting from a nontraumatic cause is a rare event, especially in infants with no underlying coagulopathy. In our patients the subdural hematoma was found incidentally, during an evaluation for increased head circumference. All infants were healthy and completely asymptomatic at presentation with no history of head trauma. There was no history of an underlying coagulopathy and no findings by history, physical examination, or x-rays to suggest child abuse. The MRI findings of bilateral prominent extracerebral CSF spaces suggested the diagnosis of external hydrocephalus in our patients.

External hydrocephalus is regarded as a benign condition, consisting of bilateral decreased densities over the frontal convexities, prominent cerebral sulci, slightly enlarged ventricles, and normal brain size [8].

The infants are usually asymptomatic and are being studied with a CT or a MRI as part of an evaluation of the large head. The syndrome is benign and the radiologic findings usually resolve spontaneously by the age of 2 years [9].

The combination of external hydrocephalus and frontal subdural hematoma in an otherwise asymptomatic infant is rare and only few cases were reported [10,11]. Azais and Echenne, in a series of 41 infants with external hydrocephalus, describe three in which subdural hematoma was found as an incidental finding without any symptoms of high intracranial pressure. Two of those infants had mild axial hypotonia at the time of presentation, but all of them had a normal neurologic examination and normal development in further follow-up [12]. The question of whether the external hydrocephalus is responsible for the subdural hematoma or is secondary to it is often raised. In a series of patients with enlarged extracerebral spaces and associated subdural hematomas by Kapila et al., radionuclide cisternography results were not indicative of communicating hydrocephalus therefore suggesting that the enlarged subarachnoid spaces were the predisposing factor [13]. At least one of our patients (Case 3) had a previous head CT revealing external hydrocephalus only without the subdural hematoma. These findings may suggest that in this group of infants stretching of the bridging veins probably occur inside the enlarged subarachnoid spaces, and the absence of an appropriate support makes them vulnerable to bleed either spontaneously or with only a minimal trauma. Recently a theoretical mathematical model of the cranial vault by Papasian and Frim, produced a relationship between venous stretch and the width of the extra-axial spaces, supporting the predisposition towards extra-axial bleeding with only minimal trauma in infants with benign external hydrocephalus [14].

The prognosis of this group of infants that are otherwise healthy is usually better compared with other cases with subdural hemorrhage [13]. In most infants the hematoma will resolve spontaneously. In two of our patients the hematoma resolved spontaneously, and the infants remain neurologically intact and achieved normal developmental milestones in 1-year follow-up. One infant (Case 3), although completely asymptomatic, was sent for neurosurgical consultation and had a surgical drainage of the hematoma. He also had no neurologic sequel and continued to achieve normal developmental milestones.

We suggest that in infants with external hydrocephalus, the enlarged subarachnoid spaces and the secondary stretching of the bridging veins may be predisposed to the development of subdural hematoma with minimal or no trauma. Child abuse should always be kept in mind, and should be ruled out in infants with subdural hematoma, but in the specific context of young infants with external hydrocephalus, subdural hematoma may be spontaneous in nature with probably good prognosis.

References

Investigating subdural haemorrhage in infants

A M Kemp

When an infant or young child presents with subdural haemorrhage, the diagnostic priority is to exclude physical child abuse. A team approach should be adopted for the clinical child protection investigation. The diagnostic process is inevitably one of detective work; appropriate radiological, ophthalmological, haematological, biochemical, and postmortem investigations are discussed.

Subdural haemorrhage (SDH) arising from intentional injury is relatively common in infants, with an annual incidence figure of 21/100 000. The mortality from shaken baby syndrome is serious: 12–30% of victims die, and 40–70% of the survivors suffer from significant neurological handicap. When an infant or young child has an SDH, the diagnostic priority is to exclude physical child abuse which is the commonest cause following shaking or shaking impact injury. Research suggests that when these children are admitted to hospital, they are often incompletely investigated. Information collected at this stage forms the major component of evidence for key decision making throughout the child protection process, within civil, criminal, and compensation litigation. The consequences of missing a diagnosis of physical abuse may leave children at risk of further injury, while an incorrect diagnosis of shaken baby syndrome will have profound effects on a family unit.

There are a number of features associated with SDH that raise the probability of abuse. These include retinal haemorrhages, additional physical injuries, and a previous history of child abuse in the family.

As with any other clinical discipline, the field of child protection must be subject to the rigours of evidence based practice and national clinical guidelines. There are currently however few systematic reviews in this field and a paucity of publications concerning the overall investigation of SDH. The literature and child protection legislation support a comprehensive interagency assessment of the child and family. All young children who have a “subdural haemorrhage diagnosed on admission to hospital where there is no clear underlying medical cause or history of witnessed major accidental trauma” must have a series of essential baseline investigations. There must be a systematic interpretation process to decide whether physical child abuse has occurred or not.

Table 1 outlines the causes of SDH in infancy and early childhood. Findings must be interpreted in the context of the different causes; all have their own specific physical signs or diagnostic markers.

TEAM APPROACH

The clinical child protection investigation should be led by a paediatrician with expertise in the field. Optimal care of the sick child depends on a partnership between the clinical team and the caregivers, and there is often an anxiety that this will be jeopardised by invoking child protection procedures. A clinical specialist in the field can address the situation with the required level of objectivity and work with local authority child protection teams. When the child is admitted to a tertiary centre or adult centred neurosurgery unit, child protection procedures are often not initiated at a local level.

Infants suffering from SDH are commonly seen in paediatric neurology units; however, the general paediatrician will be involved with a case less frequently. The clinical expertise of the paediatric neurologist or paediatric neurosurgeon will therefore be invaluable in the discussion and management of all cases.

The Local Authority Child Protection Team must be involved early to undertake preliminary investigations and exclude any previous concerns of child abuse within the family unit. Early involvement of the police will identify relevant criminal records of the child’s carers and ensure that any forensic investigation can be initiated as soon as possible.

HISTORY AND EXAMINATION

An SDH is usually diagnosed in an infant who presents as an acute hospital admission where child abuse is not the primary presenting complaint. The clinical presentation is variable in terms of severity, symptoms, and signs: There is a rarely a straightforward history of events preceding the diagnosis. Frequently the carers do not identify trauma as the most likely cause of the child’s symptoms and when asked for a possible cause, may propose various explanations from the days prior to admission that have little relevance.

Pediatricians are unused to conducting “forensic style” inquiries and are reminded that they can contaminate evidence by overzealous examination. Open ended questions will uncover the dynamics of any traumatic events described; such as the height and forces of any alleged fall or impact, the part of the body involved, how hard the surface was, and what was the child’s response after the injury. The clinician needs this..
### Table 1  Recognised causes of subdural haemorrhage in infants and young children

<table>
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<th>Cause of SDH</th>
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| Intentional injury                                                           | The commonest cause of SDH<sup>4</sup>  
  Shaken baby syndrome                                                        | SDH is commonly associated with other injuries and retinal haemorrhages.  
  However shaken baby syndrome can cause isolated SDH or isolated retinal  
  haemorrhages.                                                                 |
| Non-intentional injury<sup>27</sup>                                          | Minor household falls rarely cause SDH.  
  SDH has been described in more serious falls and in whiplash injury<sup>27</sup>  
  Retinal haemorrhages are only associated with severe occipital injury<sup>7</sup> |
| Neurosurgical complications                                                  | SDH is commonly reported as a postoperative complication of neurosurgery                                                             |
| Perinatal                                                                    | SDH infrequently reported on fetal ultrasound scans  
  Traumatic labour<sup>29</sup>                                               | SDH can follow traumatic delivery and be associated with retinal haemorrhage                                                         |
| Cranial malformations                                                          | Spontaneous bleeding from vascular malformations  
  Aneurysms<sup>10</sup>                                                       | Spontaneous bleeding from vascular malformations.  
  Aneurysms<sup>10</sup>                                                       | Less serious trauma can result in SDH when aneurysms are present.  
  Both are unlikely to be associated with retinal haemorrhages unless intracranial  
  pressure is raised.                                                                 |
| Cerebral infections                                                            | Postinfarct subdural effusions are seen.  
  Meningiomas<sup>12,23</sup>                                                   | These can be associated with retinal haemorrhage<sup>7</sup>                                                                          |
| Coagulation and haematological disorders                                      | Diagnosis will be excluded on coagulation  
  Leukopenia<sup>24</sup>                                                        | Diagnosis will be excluded on coagulation.  
  Sickle-cell anaemia<sup>23</sup>                                               | and haematological investigations. All may be associated with retinal haemorrhages<sup>7</sup> |
| Disseminated intravascular coagulation                                         |  
  Haemophilia<sup>25</sup>                                                       |                                                                                           |
| von Willibrand's disease                                                       |  
  Hereditary haemorrhagic disease of the newborn<sup>27</sup>                    |                                                                                           |
| Idiopathic thrombocytopenia purpura<sup>13</sup>                              |  
  Metabolic disorders                                                           |                                                                                           |
| Glutaric aciduria<sup>15</sup>                                               | Associated with widening of subdural  
  Golecetassaemia<sup>22</sup>                                                    | space that can result in SDH due to stretching and rupture of subdural vessels.  
  Case reports describe associated retinal haemorrhages in both conditions<sup>14,49</sup> |
| Biochemical disorder                                                          | Hypernatraemia<sup>44</sup>                                                     | SDH described in association with salt poisoning, hypernatraemic dehydration  
  Hypernatraemia<sup>44</sup>                                                   | Hypernatraemia may also be a complication of the intracranial trauma. |

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level of information to determine whether the proposed explanation is plausible.<sup>4</sup> The number of different explanations and variation in detail from one raconteur to another must be recorded. This detail must complement a full paediatric clinical history and examination that can be overlooked during the intensive management of the sick child. Any additional signs of physical abuse should be recorded in detail, injuries measured, and clinical photographs obtained.

**RADIOLOGY**

Acute haemorrhage is more easily seen on cranial computed tomography (CT) than on magnetic resonance imaging (MRI). A CT scan is more readily obtained for the acutely unwell child and is more widely available.<sup>4</sup> However, small quantities of subdural blood may be invisible on CT and it can be difficult to differentiate from fluid in the subarachnoid space. Chronic SDH may therefore look very similar to benign enlargement of the subarachnoid space which can be seen in CT scans of infants.

MRI will identify small areas of SDH and can visualise blood in positions that are not well seen on CT scan, such as the floor of the middle and posterior cranial fossae. It is also more sensitive than CT in identifying underlying parenchymal brain injury from shearing forces sustained during shaking.<sup>4</sup> MRI can detect SDH of different ages; chronic (low attenuation) subdural collections on CT often have different signal intensities on MRI which allow differentiation in time. This is of great importance when assessing the likelihood of

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repeated abuse; however, it does not follow that MRI enables accurate estimates of the ages of these bleeds.

All investigations requested should be accompanied by clear clinical details and relevant questions to the radiologist. The sequence of investigations that best identifies the effects of the injury and its sequelae, as well as detecting signs of any previous episode of abuse is therefore: a CT scan as the first line investigation, followed by an MRI within the first 7–10 days, repeated some 14 days later. Some tertiary centres, where there is a wider availability of MRI and expertise, are relying purely on sequential MRI as the imaging of choice. In expert hands cranial ultrasound may be incorporated as an additional diagnostic tool to monitor findings; however, paediatricians and general radiologists should not rely on cranial ultrasound as an exclusive investigation to identify or exclude an SDH.

Ideally a neuroradiologist with expertise in the field should report these investigations promptly. Many will be performed in a centre without such onsite support, when it may be necessary to ask for an opinion from the local neuroradiology centre. Some hospitals have image links to regional centres to facilitate this process.

SKELETAL RADIOLOGY
Skeletal fractures are common in these children.*** Rib fractures are consistent with a squeezing injury, where the infant is grasped around the chest and shaken.*** Cervical skull fractures support a shaking impact injury.*** Long bone and metaphyseal fractures need to be excluded. The latter are seen in association with shaking from the indirect acceleration deceleration forces to the fragile growing plate of the long bones or from forceful pulling or twisting of the limbs.*** An early skeletal survey should be undertaken when the child is clinically stable and repeated at 10–14 days.*** In practice the repeat skeletal survey is often omitted; however, follow up radiology may reveal previously unidentified fractures and enable more accurate dating of those already identified. Some centres perform an isotope bone scan in conjunction with the initial skeletal survey, which can identify hot spots from the early fracture healing process or subperiosteal haemorrhage. Fractures at the growing points of long bones are, however, difficult to identify on isotope bone scan.

OPHTHALMOLOGY EXAMINATION
Ophthalmological examination of the infant's eye is difficult and paediatricians often have a poor success rate. Retinal haemorrhages are characteristically at the periphery of the retina and are difficult to see with direct ophthalmoscopy. All children should have a retinal examination of both eyes, performed by an ophthalmologist with paediatric experience. Examination should use the indirect method after dilatation of the pupils as recommended by the working party of the Royal College of Ophthalmologists.***

HAEMATOLOGY AND BIOCHEMISTRY
These children often have a low haemoglobin on admission or within the first 24 hours, which may reflect the timing and extent of bleeding. A full blood count and coagulation studies will indicate blood loss, coagulation, and haematological abnormalities. Blood cultures, urea and electrolytes, and liver function tests are important to screen for infection, associated biochemical disorders, and possible intra-abdominal injury. Screening for rarer metabolic conditions is not routinely recommended. SDH in association with glutaric aciduria is seen in cases with frontotemporal atrophy.*** If the radiological findings support this possibility, glutaric aciduria should be excluded in consultation with a paediatrician who specialises in metabolic conditions.

STRATEGY MEETING
There must be an early strategy meeting with all agencies to discuss the findings and come to a joint decision about the probability of child abuse. Social services will invoke section 47 of the Children Act and initiate a child protection investigation in the majority of cases, while making provision for the immediate safety of the child and siblings.

In the cases where the cause of the SDH has not become evident and physical abuse is unlikely, consultation with tertiary specialists is recommended to exclude rarer causes.

AT POSTMORTEM EXAMINATION
Physical child abuse should be considered in any infant who dies unexpectedly. In an ideal world all such postmortem investigations should be undertaken by a paediatric pathologist in consultation with a forensic pathologist and according to nationally recommended protocols.*** When SDH is diagnosed at postmortem investigation, the diagnosis needs to be approached with the same rigour and multiagency involvement. Investigations must include a full dissection of the eyes and a complete radiological skeletal survey.*** Detailed histopathology techniques can be employed to identify diffuse axonal injury and identify the degree of brain repair in order to give an idea of the timing of injury.

WHEN TO PERFORM CT
We know that some children with SDH present with relatively mild symptoms.*** It is therefore important that paediatricians maintain a low threshold for considering this diagnosis. Many children will have a lumbar puncture as the first investigation to exclude meningitis; if this shows evidence of uniform bleeding or subarachnoid haematoma, it must be followed up with a CT scan to exclude intracerebral bleeding.

In the investigation of any child under the age of 2 years who is referred under the Child Protection Procedures, consideration should be given to performing a CT scan. This should be mandatory if the child has retinal haemorrhages, unexplained neurological findings, or an increasing head circumference, and strongly recommended in any infant with bone fractures or non-accidental bruising, or under 6 months old.

QUESTIONS IN COURT
As well as endorsing the value of good quality evidence, it is important to recognise the limitations of the evidence at various points in the process. This is never more relevant than in court, where clinicians give their opinion as expert witnesses. Questions that commonly arise concern the timing of the injury and the mechanism and forces needed to cause an SDH. The evidence to contribute to this debate is limited.

Opinion about the age of SDH is based on MRI imaging that can only suggest whether bleeds have occurred within the previous week or are older, resolving haemorrhages.*** Precise dating of associated long bone fractures is not possible, but an approximate time band can often be given according to the degree of healing evident on x-ray examination.*** The colour changes in retinal haemorrhages and bruises depend on the amount of bleeding into the retina or subcutaneous and surrounding tissue respectively, and a variable rate of resolution. Both have a red appearance when acute and a range of colour change when older.*** Severe retinal haemorrhages may take months to clear, milder ones resolving within weeks.*** Spectrophotometry appearance of cerebrospinal fluid after an intracranial bleed, the degree of cerebral oedema, and details of autopsy findings can contribute to the timing of injury.*** Discussion of the clinical features and the chronology of events, identified in the police and social assessment, often builds a more accurate forensic picture; however, accurate timing of the injury is rarely possible.
Table 2  Essential baseline assessment of an infant or young child with SDH

| Multidisciplinary team members | Paediatrician with expertise in child protection  
|                              | Paediatric neurologist and/or neurosurgeon  
|                              | Neuropathologist  
|                              | Ophthalmologist  
|                              | Area child protection team social worker and police  

Clinical history  
- Full paediatric case history  
- Full documentation of all possible explanations for injury  

Social and police history  
- Identify any previous child protection concerns, relevant criminal record of carers  

Examination  
- Thorough general examination  
- Documentation and clinical photographs of coexisting injury  
- Monitor head circumference  

Ophthalmology  
- Ophthalmologist to examine both eyes using indirect ophthalmoscope through dilated pupils  

Radiology  
- Initial cranial CT scan  
- Repeat neuroimaging at 7 and 14 days (MRI scan preferable)  
- Follow-up neuroimaging with neuroradiologist  
- Full skeletal survey: repeat imaging at 10-14 days  

Sedation  
- Full blood count repeated over first 24-48 hours  
- Coagulation screen  
- Urea and electrolytes, liver function tests, blood cultures  

Early strategy meeting of all agencies involved to come to a joint decision about the likely cause of SDH and appropriate line of management.

| Possible outcomes | Action  
|-------------------|--------  
| Likely physical child abuse | Social services will invoke section 47 of Children Act, initiate a child protection investigation and make provision for the immediate safety of the child and siblings  
| Medical cause of SDH identified | No further child protection concerns: continue medical management  
| Physical child abuse unlikely, cause of SDH unknown | Further clinical investigation in consultation with tertiary specialists to fully exclude all other causes of SDH

Knowledge of the mechanism and the forces required to elicit SDH are based on evidence from small case studies, where perpetrators have admitted shaking the baby,2 study of the clinical features of domestic and serious head injuries in children,21,22 old animal based studies where monkeys have been shaken,23 and biomechanical modelling experiments.24 These studies convince us that SDH occurs after violent whipping (acceleration-deceleration injury), but none as yet can identify the least force required for such an injury.

The clinical spectrum of these cases varies from children with multiple injuries as a result of a severe degree of violence to a child with an isolated SDH. There are accounts of babies shaken by carers with postnatal depression who cannot cope with the crying, and carers who have shaken to resuscitate an apnoeic baby. It is clear that even when the full clinical picture is evident, difficult decisions need to be made as to the intent of the assailant to harm the child.

CONCLUSIONS

The diagnostic process is one of detective work from the outset of an infant who presents with a variety of symptoms and rarely any clear history of cause. A consistent approach to the investigation, terminology, and interpretation of findings will improve the quality and accuracy of the diagnosis and management of an infant with SDH (see table 2). This must be complimented by seamless interagency cooperation with clear lines of communication.

There is evidence that clinicians are reluctant to consider a diagnosis of child abuse and often delay or fail to make an early referral to the child protection agencies.20 This can put the child and siblings at risk of further abuse, can obviate the collection of police evidence, and hinder the diagnostic process. In accordance with the Children Act 1989,21 any one who has concern that a child is suffering from significant harm should refer the matter to social services.

Research in this field is expanding and will inevitably lead to revised recommendations over time as our understanding of the condition improves.

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