CME Article

Bruises, blood coagulation tests and the battered child syndrome

Lee A C W

ABSTRACT

Cutaneous bruises are a common symptom and a sign of injury and blood coagulation disorders in childhood. A carefully-taken history, coupled with a thorough physical examination, would lead to the diagnosis, or guide the clinician to the necessary laboratory investigations. Most children suffering from non-accidental injury can have their diagnosis established on clinical grounds alone and do not require laboratory investigation. An initial screening with full blood counts, prothrombin time and activated partial thromboplastin time will be adequate in most cases if laboratory investigation is indicated, but the clinician must be aware of the limitations of these tests. The finding of an abnormal coagulation test does not exclude child abuse as it can be a consequence of maltreatment, or the two conditions may coexist. Whenever necessary, the opinion of a haematologist should be sought in order to obtain an accurate diagnosis, which is essential for subsequent management and the prevention of further injury in the case of child abuse.

Keywords: battered child syndrome, blood coagulation disorders, blood coagulation tests, child abuse, haematological diagnostic errors

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Table I. Important clues from the history and physical examination for the recognition of child abuse.

Delay in seeking medical attention.
Inconsistent or absent account of the trauma.
Pattern of bruises incompatible with the alleged mode of injury.
Timing of injury incompatible with the morphology of bruises.
Multiple bruises in the non-ambulating child.
Telltale signs of bruises indicative of an inflicted nature.

Table II. Important features suggestive of systemic bleeding tendency.

During infancy:
Delayed separation of umbilical cord or excessive bleeding afterwards.
Excessive or prolonged swelling after immunisation.
Exclusive breastfeeding and lack of vitamin K supplementation.
Prolonged bleeding after circumcision.
Throughout childhood:
Prolonged bleeding after surgery, injury or wound suturing.
Unexplained muscle or joint swellings.
Recurrent epistaxis or gum bleeding.
Recurrent bloody diarrhoea or haematemesis.
Family history of bleeding disorders.
During adolescence:
Menorrhagia resulting in significant anaemia.

INTRODUCTION

Child abuse and neglect have been recognised as a global detriment to child health and development. According to the World Health Organisation (WHO), an estimated 31,000 deaths in children were attributed to homicide in the year 2002.[1] Mortality figures represent only a tiny portion of the total burden of child maltreatment cases. Child abuse and exploitation are believed to be under-reported in the region of Southeast Asia. Many cases are concealed because child maltreatment is deeply rooted in the cultural, economic and sociocultural environment.[2] Nonetheless, the health and social consequences of child abuse are more than just death and physical injury. Victimisation during childhood is strongly associated with risk-taking behaviours later in life, such as perpetuation of violent behaviour, smoking, alcoholism, high-risk sexual behaviours, early and unintended pregnancies, depression and juvenile delinquencies. Child abuse is therefore an adverse childhood experience that is costly to the individual child, and the society as a whole.[1,3]

Among the four types of child abuse recognised by the 1999 WHO Consultation on Child Abuse Prevention, physical abuse is often the most discernable form of maltreatment.[4] Clinicians are often consulted or brought to the attention of the various forms of injuries and bruises in children. It is imperative that clinicians should be alert to the possibility of child abuse, to be able to differentiate abusive from unintentional injuries, and to recognise bleeding tendencies in children where specific investigations and treatment are needed.[5,6] On the other hand, the evaluative process should be conducted in a sensitive manner to avoid additional distress to the children if they have been traumatised. Unfortunately, evidence-based guidelines in this area are lacking. The following discussion will focus on the evaluation of the bruised child, and the diagnostic pitfalls in the setting of suspected non-accidental injury such that the affected child can be assessed in a balanced and practical approach.

Children's Haematology & Cancer Centre, East Shore Medical Centre, #05-01, 319 Joo Chiat Place, Singapore 427989

Lee A C W, MBBS
Consultant

Correspondence to:
Dr Anselm Lee Chi-wai
Tel: (65) 6340 8610
Fax: (65) 6344 0117
Email: acw_lee@parkway.sg

Table III. Reported cases of bleeding disorders mistaken as child abuse.

Gender/age of child	Underlying disease	Clinical manifestations	Reference
Male/10 mth	Haemophilia A	Widespread bruises of different ages; old fracture of clavicle	21
Male/4 mth	Vitamin K deficiency & cystic fibrosis	Recurrent bruises, petechiae, failure to thrive and anaemia	
Male/1 yr	Vitamin K deficiency & cystic fibrosis	Generalised bruises	
Female/2 yr	Acute lymphoblastic leukaemia	Recurrent bruises for one month; died with widespread leukaemic infiltration and haemoglobin of 1.8 g/dL	
Male/4 yr	Meningitis & disseminated intravascular coagulation	Multiple bruises, fever, lethargy and death	
Female/2 yr	Meningitis & disseminated intravascular coagulation	Bruises on cheek and thigh, fever and death	
Male/3 yr	Haemophilia A	Multiple bruises	18
Not mentioned	Three cases of immune thrombocytopenic purpura	Widespread bruises	
Not mentioned	Haemorrhagic disease of newborn	Not mentioned	
Male/10 mth	Haemophilia A	Recurrent spontaneous bruises and ecchymosis, epistaxis, and oral mucosal bleeding	19
Female/9 mth	Glanzmann's thrombasthenia	Recurrent bruises and epistaxis	
Female/1 yr 7 mth	Acquired inhibitors to factors II, VIII and IX	Spontaneous bruises and ecchymosis for two weeks after a diarrhoeal illness	20
Male/5 yr	Immune thrombocytopenic purpura	Three-day history of unexplained bruises	22
Female/2 yr	Immune thrombocytopenic purpura	Two-day history of unexplained bruises	
Male/8 mth	Haemophilia B	Insidious onset of unexplained bruises, especially when the child attempted to walk	
Male/14 mth	Factor XIII deficiency	Delayed cord separation with excessive bleeding; multiple intracranial haemorrhages	23

THE MEDICAL HISTORY

Taking a reliable medical and social history in the setting of suspected child abuse is seldom simple or straightforward. Because of the fear of the consequences, the accompanying caretaker is often unwilling to disclose the manner in which the child was injured. For the same reason, but not the same kind of fear, the child victim will find it difficult to tell the truth when the caretaker is around. In some cases, the child is forced to offer a fabricated history as an explanation of his or her own illness. In other cases, the caretaker alleges that the child has a tendency to bruise easily. Yet, in some other cases, especially in young toddlers or infants, no reasons will be given at all. However, the medical history remains a powerful tool to pick up important clues for the recognition or even the diagnosis of child maltreatment (Table I), to differentiate the child with a genuine bleeding tendency (Table II), and to guide subsequent laboratory investigations if necessary.[7] In order to accomplish these tasks, the clinician must approach the child and the family in a systematic and tactful manner. A multidisciplinary team support would be most helpful if this is available.

When a bleeding diathesis is suspected, the child should be evaluated with respect to his or her personal history of bleeding as well as the bleeding history in the family.[5,6] The bleeding history should include: (1) acute or chronic or recurrent pattern of bleeding, (2) age of onset, (3) recent infection or drug exposure, (4) morphology of cutaneous bleeding, (5) localised or multiple sites of bleeding, (6) involvement of mucous membranes, muscles, joints, or other internal organs, and (7) consequences of the bleeding, such as anaemia or need for transfusion treatment. For instance, children with hereditary bleeding disorders often present early, usually by the time they start ambulating, and there may be a positive family history. Boys with haemophilia typically present with intra-articular or intramuscular bleeding. Immune thrombocytopenic purpura is characterised by petechiae or pin-point haemorrhages in the skin and mucous membranes. The onset is often abrupt and there may be a preceding account of infection or vaccination. Menorrhagia is a particularly troublesome symptom in post-pubertal girls with systemic bleeding disorders. On the other hand, a clinically significant bleeding diathesis can be excluded if the child had undergone surgery, such as tonsillectomy or appendectomy, without excessive, prolonged or delayed haemorrhage.

When an apparently traumatic event is given to account for the child's bruises, the clinician should enquire into the details of the trauma. When did the "accident" occur? Where and how did it happen? Who else was present? What did the caretaker do afterwards? It is quite useful for the clinician to ask the caretaker a second time about the incident a day or two later. When the account of injury is fabricated, the caretaker may forget some of the details and come up with a different version. On other

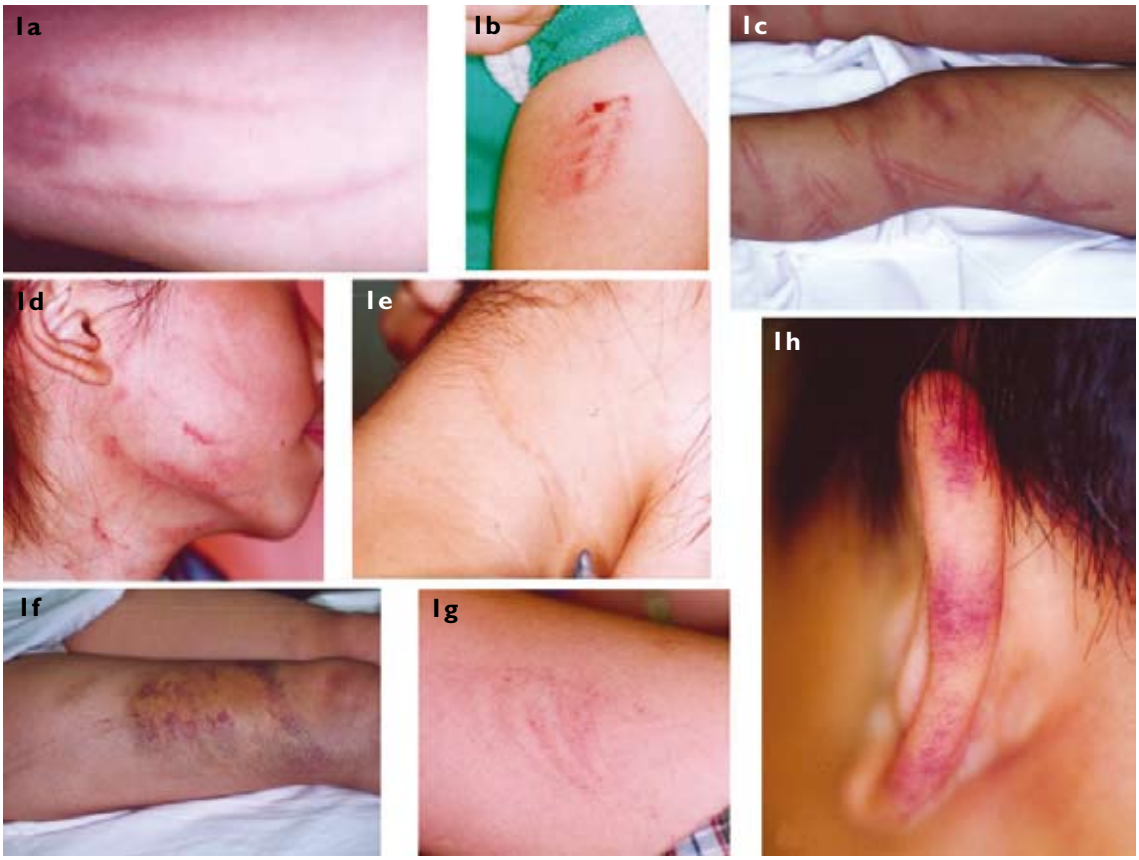

Fig. 1 Photographs of cutaneous bruises that are highly indicative of child physical abuse: (a) Characteristic loop-shaped bruise from beating with a coat hanger. (b) Spanking marks caused by a back-scratcher. (c) Multiple spanking marks caused by a rattan cane. (d) Slap marks caused by blows on the cheek. (e) Ligature mark around the neck. (f) Multiple bruises from beating with a metal pipe. (g) Imprint from beating with a slipper. (h) Pinch marks on the pinna.

occasions, the caretaker may try to "correct" the earlier account of injury when they suspect the first account was not convincing enough. In older children who can verbalise, the clinician should create an opportunity to talk to them when their caretakers are not around. Given a non-threatening environment, supported by caring people, and sometimes with a gesture to suggest that the first account of injury is not credible, the child may be able to articulate how the actual injury happened.[8]

THE PHYSICAL EXAMINATION

The child should be evaluated with respect to his or her growth parameters. Physically-abused children are often normal in terms of growth and nutritional status, unless they are victims of neglect as well. The sites and morphology of the bleeding should be carefully documented, and associated injuries should be looked for. Cutaneous ecchymosis and intra-articular bleeding are typically seen in patients with haemophilia. Deformity of the joints and muscle wasting are common accompanying signs in older haemophilic children. Children with thrombocytopenia or congenital thrombocytopathy often present with mucocutaneous bleeding and petechial haemorrhages. The latter bleeding is often reproduced in the arm after a tourniquet is tied for venipuncture, or seen in the face after vigorous crying.

Disorders in connective tissue is an uncommon cause of easy bruising and may rarely be confused with child abuse.[9,10] The vascular fragility that gives rise to cutaneous bruises is often found along with generalised connective tissue fragility, skin hyperextensibility, joint hypermobility and paper-tissue scars. Generalised petechial bleeding associated with gingival hypertrophy and bleeding is rarely seen in children with vitamin C deficiency, although a recent case reported an autistic child who had been given a highly-restricted diet.[11] A perifollicular pattern of cutaneous bleeding is characteristic in scurvy.

On the other hand, when bruises occur as a result of inflicted injury, the bruises may conform to the shape of the injuring instrument (Fig. 1). Slapping and beating with a stick or rod leave characteristic marks that even non-medically trained personnel can identify. Bruises left by beating with a belt or strap, belt buckle, coat hanger, electric cord, rubber hose, the soles of a shoe or sandal often leave typical patterns of bruises that can be matched to the shape of the instrument. A careful search and documentation of such injuries can provide

powerful evidence to child abuse. Additional findings, such as injury to the labial frenum, traumatic rupture of the tympanic membrane and fractures, can further support the diagnosis of child abuse in uncertain cases.

DATING BRUISES

While cutaneous bruises show sequential colour changes as the extravasated blood in the subcutaneous region is gradually decomposed and reabsorbed, the timing that each of the different discolouration takes place is highly variable. A purplish-reddish discolouration is generally a sign of a "fresh" bruise which can be seen as long as seven days after the injury. The appearance of a greenish-yellowish discolouration generally signifies an "old" bruise, but can be seen as early as 24 hours after the injury.[12] It is not surprising when a group of 50 children presented with accidental bruises to an emergency department, the accuracy of dating of the bruises by a group of 63 clinicians ranged from 0% to 100%.[13] Thus, under most circumstances, dating a bruise is an inaccurate practice. Bleeding under the mucous membranes does not undergo colour changes, and hence can never be dated.

BLOOD COAGULATION STUDIES

The majority of children undergoing evaluation of suspected child abuse do not require laboratory investigations, as the diagnosis can be established by a carefully-taken history and thorough physical examination and documentation. Of the 320 children admitted consecutively for evaluation of child abuse in Hong Kong, only 51 (16%) required blood coagulation studies while maltreatment was ascertained in 74% of them.[7] Blood coagulation tests are indicated only when a bleeding diathesis is suspected on clinical grounds, or when a pattern of bleeding remained unexplained after initial evaluation. It is unfortunate that in some jurisdictions, blood tests have become a legal necessity and this has been viewed as a form of "abuse".[6]

In general, full blood counts, prothrombin time (PT) and activated partial thromboplastin time (APPT) measurements are commonly used as screening tests. Tests for bleeding time may be required if a qualitative platelet defect is suspected. The need for more specific tests, such as bone marrow examination when malignancy is suspected, platelet function studies, and measurement of specific coagulation factor activities, will be determined by the findings of the screening tests.

Rarely, children with hereditary bleeding disorders may present with normal platelet counts, coagulation screens and bleeding time. Factor XIII deficiency classically presents as prolonged bleeding from the umbilical cord, delayed bleeding from injury and intracranial bleeding.[14] Its presence may be suspected when clot stability cannot be sustained in urea or acetic acid solution. Patients with hereditary disorders in fibrinolysis such as α_2-plasmin inhibitor (antiplasmin) deficiency and plasminogen activator inhibitor-1 deficiency may be clinically indistinguishable from haemophilia.[15] Specific functional assays are needed to confirm their diagnosis.

With this clinically-based evaluative approach and selective use of laboratory tests in 16% of the cases, the author has been able to ascertain 568 (79%) of 720 children suspected of child physical abuse in Hong Kong,[4] while diagnosing factor II deficiency and immune thrombocytopenic purpura, respectively, in two of the children.[7] The diagnosis of inflicted head injury (or the shaken baby syndrome) with successful prosecution can be made without exhaustive laboratory investigations to exclude every kind of known haemorrhagic disorders.[16,17]

PITFALLS IN DIAGNOSIS

Diagnostic errors may occur when a child with a genuine bleeding disorder is mistaken for child abuse, or when maltreatment is missed in the child with coexisting bleeding diathesis. Children with bleeding disorders may be initially thought to be victims of maltreatment (Table III), but misdiagnosis is extremely rare when the patients are evaluated in a systematic manner. Of the 2,578 cases evaluated by the child abuse team in Leeds, only five (0.2%) children were found to have coagulation disorders, including immune thrombocytopenic purpura, haemophilia A and vitamin K deficiency bleeding.[18] Case reports of Glanzmann's thrombasthenia, haemophilia A, and acquired haemophilia have been described in the literature when an initial diagnosis of child abuse was erroneously made.[19,20] However, the correct diagnoses were made after careful clinical evaluation and timely laboratory tests.

In a review of conditions mistaken for child abuse, Bays mentioned seven cases of "occult" coagulopathies, including haemophilia, acute lymphoblastic leukaemia, vitamin K deficiency bleeding (or haemorrhagic disease of newborns), and disseminated intravascular coagulation, as examples of misdiagnosis.[21] Harley also reported two cases of immune thrombocytopenic purpura and a case of haemophilia B that were initially thought to have been due to maltreatment.[22] However, re-examination of their clinical manifestations and the final diagnosis suggest that these diagnoses should not have been missed if the children had been evaluated systematically. For instance, in the most recently-reported case, a two-year-old child had been put under social scrutiny because of suspected child abuse from an incident of "unexplained" intracranial

bleeding. However, the history of excessive bleeding requiring transfusion therapy following cord removal, and a prior account of intracranial bleeding during infancy, had not been elicited properly. Although factor XIII deficiency was suspected, the diagnosis was missed because the screening test of clot solubility was done immediately after plasma transfusion treatment.[23]

It cannot be overemphasised that the finding of a coagulation abnormality does not exclude the diagnosis of child abuse. First, coagulopathy may be a consequence of physical abuse. Of the 101 children studied by Hymel et al for inflicted head injury, 54% had mild prolongation of PT and 24% had prolongation of APTT.[24] Second, coagulopathy may be part of an induced illness. Munchausen syndrome by proxy has been reported with deliberate poisoning with rodenticide (superwarfarin).[25] Third, coagulopathy may coexist with child abuse. O'Hara and Eden diagnosed platelet aggregation disorder and von Willebrand disease in two children, respectively, in whom the diagnosis of physical abuse was also evident from the history and social enquiry.[20]

CONCLUSION

Clinicians play an important role in the evaluation of children who present with bruises. It is important to diagnose children with blood coagulation disorder timely so that they can receive the appropriate and specific treatment. It is also important to identify the children who have been abused so that preventive measures can be implemented to help them and their families. A systematic approach is necessary for accurate diagnosis, and the opinion of a haematologist should be sought when there is uncertainty in the diagnosis. A high index of suspicion is required in order for child abuse not to be overlooked.

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SINGAPORE MEDICAL COUNCIL CATEGORY 3B CME PROGRAMME
Multiple Choice Questions (Code SMJ 200806A)

	True	False
Question 1. When a child is evaluated for bruises, the presence of an underlying bleeding tendency:		
(a) Can be excluded if the family history is negative.	☐	☐
(b) Can be excluded if there is a definite history of trauma.	☐	☐
(c) Should be suspected if there is a recent history of chickenpox.	☐	☐
(d) Should be suspected if there is bleeding from more than one site.	☐	☐
Question 2. The following features favour the diagnosis of haemophilia rather than immune thrombocytopenic purpura:		
(a) A positive family history of bleeding diathesis among male offspring.	☐	☐
(b) A history of prolonged swelling after intramuscular injection or vaccination.	☐	☐
(c) A history of recurrent joint pain and swelling.	☐	☐
(d) A recent history of influenza-like illness prior to the occurrence of bruises.	☐	☐
Question 3. Child abuse should be suspected when:		
(a) Multiple bruises are found in a non-ambulating child.	☐	☐
(b) There is recurrent epistaxis.	☐	☐
(c) Patterned bruises are found compatible with beating with an instrument.	☐	☐
(d) There is inconsistency in the accounts of injury.	☐	☐
Question 4. The diagnosis of non-accidental injury:		
(a) Can be excluded when an explanation for injury can be given.	☐	☐
(b) Can be established from a thoroughly taken history.	☐	☐
(c) Can be excluded when an abnormality in blood coagulation test is found.	☐	☐
(d) Can be excluded if the child has haemophilia.	☐	☐
Question 5. A three-month-old boy is seen with a prolonged swelling in his right thigh after receiving an intramuscular injection two weeks ago. He has been exclusively breastfed since birth. A bruise quickly develops in his left arm after a tourniquet is tied for blood taking.		
(a) Platelet count measurement is not necessary as intramuscular bleeding suggests coagulation disorder.	☐	☐
(b) Vitamin K deficiency should be suspected when prothrombin time, activated partial thromboplastin time, and liver function tests are abnormal.	☐	☐
(c) Excessive bleeding from the umbilical stump separation is a characteristic sign in children with haemophilia A.	☐	☐
(d) Isolated prolongation of activated partial thromboplastin time and a markedly diminished Factor IX level is characteristic in haemophilia B.	☐	☐

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