

Child Abuse or Bleeding Disorder—An Interdisciplinary Approach

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Abstract

Children with an unexplained bleeding tendency are frequently referred to a haemostaseologist for further evaluation. Careful standardized history taking and clinical evaluation should allow for distinguishing bleeds after minor injury and trauma which are very common in all children. However, in two groups of children bleeding symptoms may be more significant than expected: those with an underlying coagulation disorder and those who have been subjected to physical child abuse. The coexistence of child abuse and a bleeding disorder must always be considered. An extended coagulation diagnostic is required if the morphology of bleedings is not clearly suspicious for child abuse and in the absence of typical concomitant injuries, e.g., bone fractures. An interdisciplinary approach involving a forensic pathologist and a paediatric haemostaseologist for assessment of bleeding symptoms, the explanation of the clinical findings, and the critical evaluation of laboratory results are essential in such cases. This review is focussed on symptoms in accidental and nonaccidental injuries in children assisting haemostaseologists in decision making in cases of child protection issues.

Keywords

- ▶ child abuse
- ▶ injuries
- ▶ bleeding symptoms
- ▶ haemostasis testing

Zusammenfassung

Kinder mit unklarer Blutungsneigung werden häufig zum Hämostaseologen zur Abklärung geschickt. Eine standardisierte Anamneseerhebung und klinische Untersuchung erlauben es zu entscheiden, ob es sich um Blutungen durch kleine Verletzungen und Traumata handelt. In der Regel treten bei 2 Gruppen von Kindern relevante Blutungen auf: Kinder mit Gerinnungsstörung und solche, die körperlich misshandelt werden. Dabei ist stets zu berücksichtigen, dass auch gleichzeitig eine Gerinnungsstörung und eine Misshandlung vorliegen kann. Eine erweiterte Gerinnungsdiagnostik ist erforderlich, wenn die Blutungsmorphe nicht eindeutig für eine Misshandlung spricht und beim Fehlen typischer Begleitverletzungen, wie Frakturen. In diesen Fällen ist ein interdisziplinäres Vorgehen unter Einbeziehung eines Rechtsmediziners und eines pädiatrischen Hämostaseologen zur Beurteilung der Symptome, den Erklärungen für das vorliegende klinische Bild und eine kritische Bewertung der Befunde der Gerinnungsdiagnostik notwendig. Diese Übersichtsarbeit fokussiert auf Symptome bei akzidentellen und nicht-akzidentellen Verletzungen und soll eine Entscheidungshilfe für Hämostaseologen in Kinderschutzfällen darstellen.

Schlüsselwörter

- ▶ Kindesmisshandlung
- ▶ Verletzungen
- ▶ Blutungssymptome
- ▶ Gerinnungsdiagnostik

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Introduction

The most easily recognizable physical signs of child abuse are seen in the skin and are often manifest as bruises.^{1,2} Although the detection of abuse is vital, the physicians involved in such cases face the problems of potential alternative diagnoses when confronted with injuries that are suspected of being nonaccidental. It may be difficult to distinguish abuse from other diseases and conditions that produce similar signs and symptoms. Clearly, part of the diagnostic algorithms must involve the identification and diagnosis of an underlying coagulation disorder. It must be clear that abnormal findings in the coagulation tests do not necessarily exclude abuse. Therefore, knowledge of symptoms in accidental and nonaccidental injuries is essential also for any physician in such cases.

Injuries

In case of relevant physical injuries in children, the plausibility of the given explanations by caregivers must be critically checked, considering the child's age, history, and level of activity (→Table 1). For instance, the onset of bleeding symptoms in infants younger than 1 year of age is extremely rare and such findings may therefore point towards nonaccidental injury or a severe form of a coagulation disorder.

Skin and Soft-Tissue Bleedings

Because skin and soft-tissue bleedings are major findings in child abuse, their precise assessment and documentation are crucial for the investigations of suspicious cases (→Tables 2 and 3). It is essential to consider the age-dependent mobility of the child. Small and locally limited haematomas are frequently observed in children starting at the end of the first year of life due to the progressive motor development and the increased incidence of minor traumas. These soft-tissue bleedings are typically located in prominent areas of the body such as in the area of the forehead, pretibial, elbow, and back of head. As shown in →Fig. 1, atypical bleedings on the other hand are localized on the chest, back, neck, genitals, the dorsal sides of thighs, and the forearms.^{1,5} Petechial bleedings occur more frequently in cases of child abuse than in accidental injuries.⁶

As depicted in →Table 3, the presence of the rare Ehlers–Danlos syndrome as a heterogeneous group of connective-tissue disorders should also be considered as the reason for easy bruising.

Intracranial Haemorrhage

The incidence of nonaccidental head injuries depends on age whereby mainly children of less than 3 years of age with a peak between 2 and 5 months are affected. Based on the high lethality ranging from 30 to 70%, this form of child abuse is

Table 1 Validity of physical injuries for the presence of child abuse (modified according to Herrmann 2002³ and Sorantin and Lindbichler⁴)

Physical injuries of high significance for child abuse	
Central nervous system (CNS)	<ul style="list-style-type: none"> ▪ Subdural haematomas in combination with retinal bleeding and neurologic symptoms ▪ Retinal bleeding ▪ Retinoschisis ▪ Vitreous body bleeding
Abdomen	<ul style="list-style-type: none"> ▪ Intramural duodenal haematoma ▪ Hollow organ perforations
Fractures	<ul style="list-style-type: none"> ▪ Classical metaphyseal bone fracture (children < 2 years) ▪ Rib fractures ▪ Fractures of scapula, processus spinosus, or/and sternum ▪ Fracture(s) within the first 6 months of life (premobile infants) without adequate history
Physical injuries of moderate significance for child abuse	
CNS	<ul style="list-style-type: none"> ▪ Subdural haematomas (particularly over the convexity, interhemispherical, and subarachnoidal)
Abdomen	<ul style="list-style-type: none"> ▪ Injuries of left liver lobe and/or kidneys and/or pancreas, pancreas pseudocysts
Throat–nose–ears	<ul style="list-style-type: none"> ▪ Hypopharynx perforations
Fractures	<ul style="list-style-type: none"> ▪ Multiple, particularly both-sided fractures ▪ Fractures of various ages ▪ Epiphyseolysis ▪ Vertebral body fractures ▪ Fractures on fingers, hands, and feet ▪ Complex skull fractures (particularly with accompanied intracranial injuries) ▪ Mandible fractures ▪ Periosteum alterations ▪ Fractures in infants
other	<ul style="list-style-type: none"> ▪ Relapsed apnoea (apparent life-threatening event)

Table 2 Specificity of skin and soft-tissue bleedings in child abuse (modified according to Herrmann 2002³)

High specificity	<ul style="list-style-type: none"> • Patterned haematomas (e.g., grip marks, finger marks, welts, belt, loops, and sticks); see also • Bite marks →Fig. 2
Moderate specificity	<ul style="list-style-type: none"> • Multiple haematomas • Unusual haematoma sites such as face, retroauricular, abdomen, buttocks, arms, and hands • Haematoma in infants
Low specificity	<ul style="list-style-type: none"> • Multiple haematomas in mobile toddlers on prominent parts of body; see also →Fig. 3 • Different coloured haematomas

Table 3 Differential diagnoses of skin and soft-tissue bleedings (modified according to Jaffe 1994⁷ and Herrmann et al 2010⁸)

Haematoma	Accidental haematoma	Preferentially on bony prominences (“leading edges”)
	Coagulation disorders	For example, coagulation factor deficiencies such as haemophilia, von Willebrand syndrome (VWS), thrombocytopenia and -pathy, vitamin K deficiency, accidental anticoagulant intake
	Congenital disorders and skin variations	Mongolian spot, Ehlers--Danlos syndrome
	Infections	Erythema multiforme
Purpura	Vasopathies	e.g., Schönlein–Henoch purpura
Petechiae	Increased pressure in the regions of head, neck, or thorax	Vomiting, cough attacks, excessive crying
	Accidents/violence	Near-drowning, incarcerations, electrical accidents, foreign body aspiration, compression of neck soft tissue (crapping, choking)
	Coagulation disorders	Thrombocytopenia and -pathy, VWS, sepsis with disseminated intravascular coagulation (DIC)
	Vasopathies	e.g., Schönlein-Henoch purpura
	Intoxications	e.g., Colchicine intoxication
	Others	Complicated spontaneous delivery, after cardiopulmonary reanimation

the most common nonnatural cause of death in infants and toddlers.^{9,10} Intracranial injuries with bleedings are due to a direct force effect as a result of blows or of kicks with clashing the skull on a tight surface and or by an indirect force effect mostly referred as “shaken baby syndrome.”

One distinguishes between epidural, subdural, subarachnoidal, and intraparenchymatous bleedings. The *subdural haematoma* originates mainly from trauma by tearing of bridging veins with consecutive bleeding between dura mater and arachnoidea. It occurs more frequently in the cases of nonaccidental injuries. The appearance of a subdural haematoma is more suspicious for child abuse compared with the *epidural haematoma* which is the consequence of an arterial bleeding and located between dura and cranium.^{11,12} Following drops of minor height no clinically relevant injuries including an intracranial haemorrhage (ICH) are expected.¹³ Brain lesions are often accompanied by a *subarachnoidal bleeding* which often occurs in cases of child abuse.

The *shaken baby syndrome* represents a common form of nonaccidental craniocerebral injury in newborns and

infants. It is associated with significant clinical consequences including retarded neurological development and tendency to seizures. It requires a massive bouncing back and forth shaking of the child who is kept tightly on the upper arms or the trunk.¹⁴ The mechanism of the forcible shaking leads to an uncontrolled head rotation and serious sheer forces in different intracranial compartments. This results in diffuse cerebral parenchyma damages and typically in the bilateral or the interhemisphere gap located subdural bleedings (see also **→Fig. 4**).^{14,15}

Pronounced retinal bleedings are highly suspicious for child abuse, especially for the act of shaking.^{16,17}

Intracranial bleedings may also occur in rare cases of inborn errors of metabolism, e.g., in glutaric aciduria of type I.^{18–20} Reports for the first manifestation of Menkes syndrome with an ICH²¹ and retinal bleedings in galactosemia exist.²²

Intracranial perinatal injuries with subdural and retinal bleeds that occur mainly in association with vacuum extraction are rarely serious^{23,24} and are typically completely absorbed approximately 4 weeks after delivery.¹⁴ History

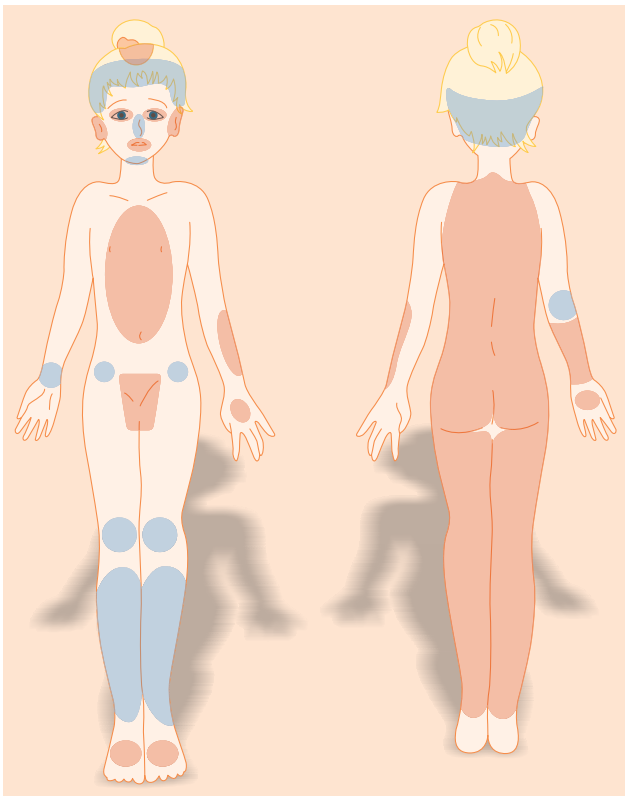


Fig. 1 Typical allocations of haematomas in injuries (in blue) and in child abuse (in red) (adapted from Knöfler et al 2014.³⁷)

of prolonged persistence of cutaneous haematoma and prolonged or severe jaundice may be an early symptom of inherited bleeding disorders.

Congenital bleeding disorders are a minor but a significant cause of ICH.²⁵ The risk of this life-threatening bleeding is highly variable and depends on the underlying disease. For instance, despite the typically very low platelet count of less

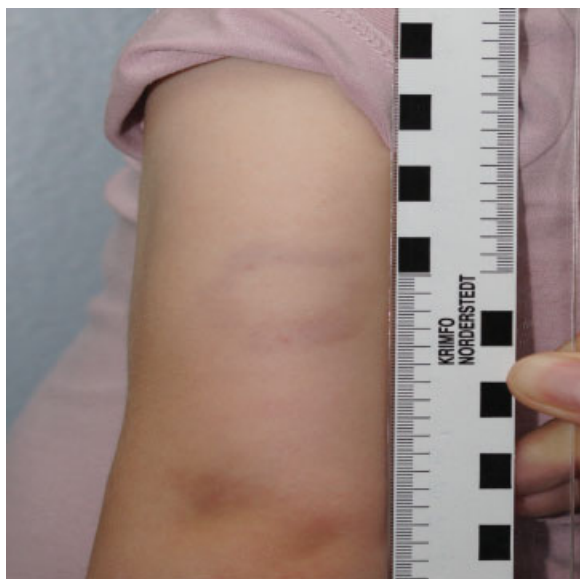


Fig. 2 Bite mark on the upper arm.



Fig. 3 Multiple pretibial haematomas in a toddler with severe haemophilia A.

than 20,000/ μ L, the risk is less than 1% in children with immune thrombocytopenia.²⁶ Most inherited platelet function disorders have a mild to moderate bleeding tendency that can never experience a severe bleeding such as ICH. However, Glanzmann's thrombasthenia as a very rare but severe form can lead to ICH. Contrastingly, in patients with severe haemophilia, ICH may occur after minor trauma but

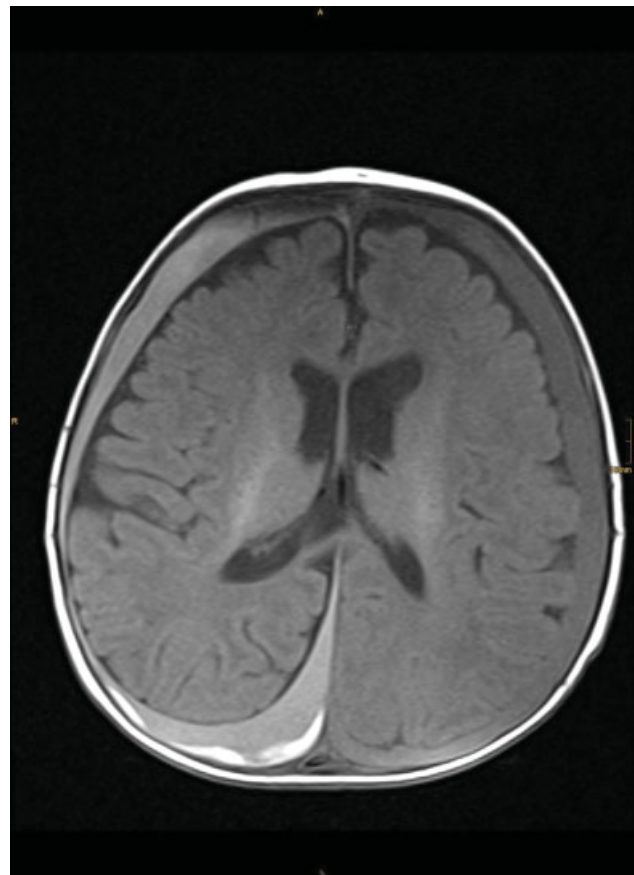


Fig. 4 Typical MRI findings in shaken baby syndrome showing bilateral subarachnoid bleedings of different ages extending into the interhemisphere gap. MRI, magnetic resonance imaging.

also spontaneously.^{27,28} More common is the ICH in patients with inherited severe factor XIII deficiency where it occurs in about one-third of these patients.²⁹

Significance of Haemostasis Testing

An unclear bleeding tendency in childhood calls for an extended coagulation work-up, particularly if child abuse is suspected and typical concomitant injuries are absent. The chosen diagnostic tests should be able to detect the presence of relatively common coagulation defects such as von Wil-

lebrand syndrome or haemophilia, but also rare diseases such as inherited thrombocytopathies. Coagulation testing in these cases should be part of an extensive diagnostic work-up depicted as a checklist shown below.

Before performing coagulation testing the evaluation of a detailed bleeding history of the child and the family members is of particular importance.^{1,30} Standardized bleeding questionnaires (ped International Society of Thrombosis and Haemostasis - Bleeding Assessment Tool (ISTH-BAT)) are especially helpful in children beyond 1 year of age.³¹

Table 4 Check list for proceedings in children with bleedings suspicious for child abuse (according to Knöfler and Schmidt 2018)³⁹

History
<ul style="list-style-type: none"> ✓ Detailed patient's history and recent situation including the injury circumstances, attendees, and behaviour of parents ✓ Critical assessment of the described course of injury event ✓ Assessment of patient's bleeding history: <ul style="list-style-type: none"> ▪ Umbilical cord bleeding and delayed cord separation ▪ Prolonged bleeding from the heel prick for blood collection testing of inborn errors of metabolism (Guthrie test) ▪ Tendency to epistaxis, haematomas (also after vaccinations), mucocutaneous bleeding, menorrhagia ▪ Peri-/postinterventional bleedings ▪ Soft-tissue and joint bleedings ▪ Gastrointestinal bleedings ✓ Intake of coagulation influencing medication and substances ✓ Assessment of family history including bleeding events, consanguinity ✓ Behaviour history: sudden emotional or behaviour modification such as sleep disturbance, regression, or aggressive behaviour
Physical examination
<ul style="list-style-type: none"> ✓ Assessment of general condition, nutritional status, and fostering condition ✓ Whole body examination including bleeding signs with exact documentation of abnormal results (location, type, and extent) using a body scheme draft and photo documentation ✓ Clinical signs of fractures ✓ If applicable anogenital examination
Apparative diagnostics/imaging
<ul style="list-style-type: none"> ✓ 0–2 years: skeleton screening using X-ray (extremities, chest, pelvis a.p., spine laterally, skull a.p. + laterally) ✓ >2 years: skeleton – clinically suspicious parts in two plains ✓ Sonography (abdomen, cranium) ✓ Retinal assessment ✓ Cerebral imaging (magnetic resonance imaging [MRI] including spinal axis, cerebral computed tomography [CT] as emergency diagnostics)
Coagulation diagnostics
<ul style="list-style-type: none"> ✓ Level 1 (preliminary coagulation assessment on-site): prothrombin time (quick), activated partial thromboplastin time (aPTT), fibrinogen, full blood cell count, and blood film ✓ Level 2 (on-site and if applicable by sending to external laboratories): von Willebrand factor antigen (VWF:Ag), collagen binding activity (VWF:CB), VWF activity, VWF-multimeric-analysis^a, ristocetin-induced platelet agglutination (RIPA)^a, coagulation factors VIII, IX, and XIII, blood group ✓ Level 3 (by referring the patient to a haemostaseologic centre): platelet function diagnostics (e.g., aggregometry, fluorescence-activated cell sorting [FACS] analysis)
Further laboratory diagnostics
<ul style="list-style-type: none"> ✓ Transaminases, pancreas enzymes, alkaline phosphatase ✓ Creatine kinase, troponin, lactate dehydrogenase, lactate, urea, creatinine ✓ Uric acid, electrolytes ✓ Blood glucose ✓ Blood gas analysis In case of suspected inborn error of metabolism: <ul style="list-style-type: none"> ✓ Copper and ceruloplasmin in serum (Menkes syndrome) ✓ Acylcarnitine profile, aminogram, homocysteine in serum (organoaciduria, aminoacidopathies) ✓ Excretion of organic acids in urine (organoaciduria) ✓ Excretion of mucopolysaccharides and oligosaccharides in urine (lysosomal storage disease) ✓ Isoelectrical focusing of transferrin and apolipoprotein CIII (apoCIII) in blood (congenital disorder of glycosylation)

^aVWF multimeric testing and RIPA only recommended if VWD type 2 suspected.

It should be taken into consideration that the global coagulation screening tests (prothrombin time [PT] and activated partial thromboplastin time [aPTT]) and the determination of blood cell count do not rule out the presence of von Willebrand disease, factor XIII deficiency, or inherited thrombocytopathies.^{32–37} However, due to the lack of clear reference ranges, especially in neonates and infants, the interpretation of some individual findings of some coagulation studies can be considerably difficult for infants below 6 to 12 months of age.³⁸ Moreover, the limited availability of coagulation tests in specialized laboratories and some preanalytical problems, such as decreased clotting factor activities due to a long sample transport to external laboratories, must also be taken into account.

Initially, coagulation diagnostics that are available on-site should be performed to implement all level 1 tests (PT, PTT, fibrinogen, and platelet counts). Level 2 coagulation tests (blood group, von Willebrand factor antigen and function, and factors VIII, IX, and XIII) cannot be done sufficiently on-site in some hospitals. Therefore, citrate plasma samples must be sent to specialized coagulation laboratories with the exception of samples for platelet function. Most platelet tests can only be performed from freshly taken venous blood samples. For this reason it is highly recommended to refer patients to a centre with expertise in bleeding disorders.

We recommend the use of a check list for proceedings in children with bleedings suspicious for child abuse (Tab. 4; according to Knöfler and Schmidt 2018).³⁹

Conclusions

Child protection is an interdisciplinary challenge. Suspicious skin bleedings may be associated with internal injuries or bone fractures. To clarify suspicious findings a child protection group consisting of paediatricians, paediatric surgeons, paediatric radiologists, forensic doctors, and social workers should discuss the diagnostic procedure together. In some cases it is necessary to involve other specialized disciplines such as ophthalmology, haemostaseology, or paediatric psychiatry. Findings should be evaluated together in a case conference.

Conflict of Interest

On behalf of all co-authors the first and the senior author declare no conflicts of interest.

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