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Evaluation for Bleeding Disorders in Suspected Child Abuse

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CLINICAL REPORT

Evaluation for Bleeding Disorders in Suspected Child Abuse

abstract

FREE

Bruising or bleeding in a child can raise the concern for child abuse. Assessing whether the findings are the result of trauma and/or whether the child has a bleeding disorder is critical. Many bleeding disorders are rare, and not every child with bruising/bleeding concerning for abuse requires an evaluation for bleeding disorders. In some instances, however, bleeding disorders can present in a manner similar to child abuse. The history and clinical evaluation can be used to determine the necessity of an evaluation for a possible bleeding disorder, and prevalence and known clinical presentations of individual bleeding disorders can be used to guide the extent of the laboratory testing. This clinical report provides guidance to pediatricians and other clinicians regarding the evaluation for bleeding disorders when child abuse is suspected. *Pediatrics* 2013;131:e1314–e1322

INTRODUCTION

Children often present for medical care with bleeding or bruising that can raise a concern for child abuse. Most commonly, this occurs with cutaneous bruises and intracranial hemorrhage (ICH), but other presentations, such as hematemesis,¹ hematochezia,² and oronasal bleeding can be caused by child abuse and/or bleeding disorders.^{3–7} When bleeding or bruising is suspicious for child abuse, careful consideration of medical and other causes is warranted. The inappropriate diagnosis of child abuse could occur,^{8–10} potentially resulting in the removal of a child from a home and/or the potential prosecution of an innocent person. Conversely, attributing an abusive injury to medical causes or accidental injury puts a child at risk for future abuse and possible death.¹¹ Laboratory evaluations should be conducted with the understanding that the presence of a bleeding disorder does not rule out abuse as the etiology for bruising or bleeding.⁹ Similarly, the presence of a history of trauma (accidental or nonaccidental) does not exclude the presence of a bleeding disorder or other medical condition. This clinical report provides guidance to pediatricians and other clinicians regarding the evaluation for bleeding disorders when child abuse is suspected (Fig 1).

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KEY WORDS

intracranial hemorrhage, inherited coagulation disorders, bruising, nonaccidental trauma

ABBREVIATIONS

aPTT—activated partial thromboplastin time
DIC—disseminated intravascular coagulation
ICH—intracranial hemorrhage
ITP—immune thrombocytopenia
PFA-100—platelet function analyzer
PT—prothrombin time
VKDB—vitamin K deficiency bleeding
VWD—von Willebrand disease

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The guidance in this report does not indicate an exclusive course of treatment or serve as a standard of medical care. Variations, taking into account individual circumstances, may be appropriate.

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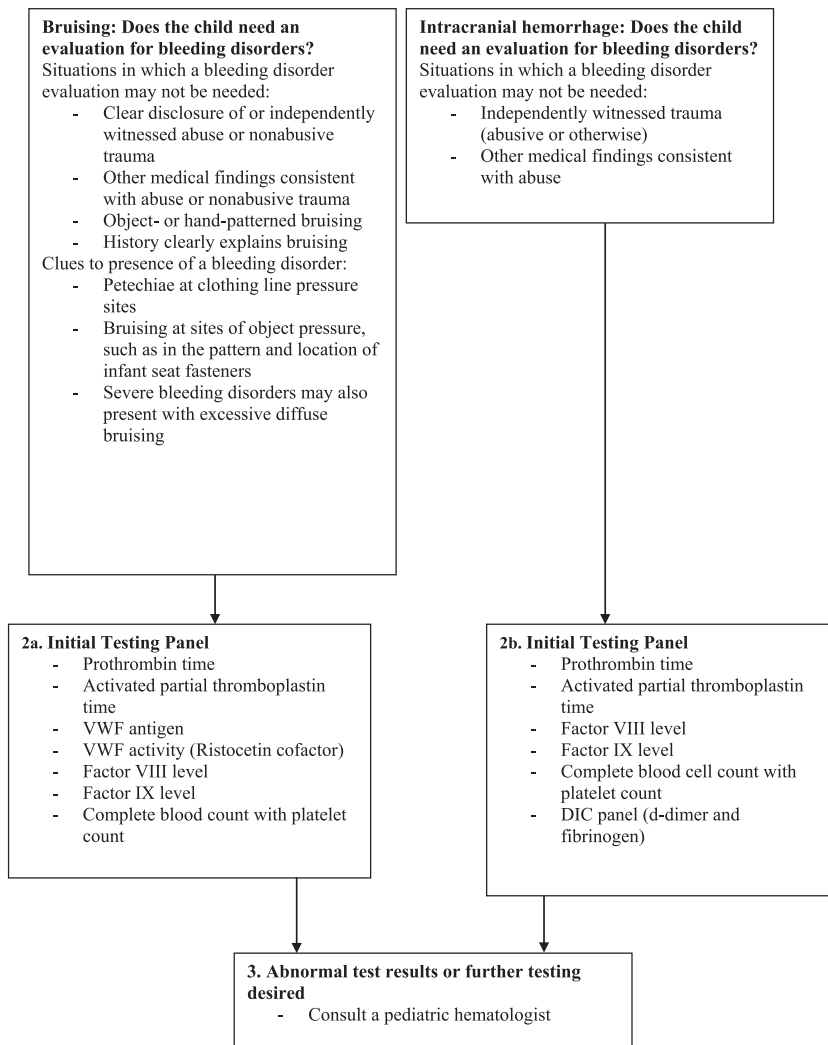


FIGURE 1
Recommended pathway for evaluation of possible bleeding disorders when child abuse is suspected. VWF, von Willebrand factor.

ASSESSING THE NEED FOR A LABORATORY EVALUATION FOR BLEEDING DISORDERS

The age and developmental capabilities of the child, history of trauma, the location and pattern of bruising, and, in the case of ICH, findings on neuroimaging should be considered when assessing children with bruising/bleeding for possible abuse.^{12–18} Additionally, a medical history of symptoms suggestive of a bleeding disorder, such as significant bleeding after a circumcision or other surgery, epistaxis, bleeding from the umbilical stump, or

excessive bleeding after dental procedures, increases the possibility of a bleeding disorder. Family history of a specific bleeding disorder or ethnicity of a population with higher rates of a certain bleeding disorder (eg, Amish) might necessitate testing for that condition. The child's medications should be documented, because certain drugs can affect the results of some tests that might be used to detect bleeding disorders, such as the platelet function analyzer (PFA-100; Siemens Healthcare Diagnostics, Tarrytown, NY) and platelet aggregation testing. Caregivers might state

that their child “bruises easily.” These statements are difficult to assess during an evaluation for possible abuse, as they can be a sign of a bleeding disorder, a reflection of the child's (fair) skin tone, or a fabrication to mask abuse. Children who are verbal and capable of providing a history should be interviewed away from potential offending caregivers, if possible. A thorough physical examination should include an evaluation of areas of bruising that have higher specificity for abuse,¹⁴ such as the buttocks, ears, and genitals.

Any bleeding disorder can cause cutaneous bruising, and sometimes this bruising can be mild, can appear in locations that are considered suspicious for abuse,¹⁹ and can appear at any age. Given the extreme rarity of some bleeding disorders, it is not reasonable to perform extensive laboratory testing for bleeding disorders in every child. In some cases, the constellation of findings, taken in conjunction with the clinical history and physical examination, can be so strongly consistent with an abusive injury that further laboratory investigation for medical conditions is not warranted. For instance, a child with a patterned slap mark who describes being hit with an open hand does not require a laboratory evaluation for a bleeding disorder.

In addition to bleeding disorders, the possibility of other medical causes of easy bruising or bleeding, such as Ehlers-Danlos syndrome, scurvy, cancer and other infiltrative disorders, glutaric aciduria, and arteriovenous malformations, should be assessed, as should a history of use of any medications or alternative therapies that may increase bleeding/bruising. Comprehensive descriptions of medical conditions that could be confused with child abuse and alternative therapies that may predispose to

bleeding/bruising are beyond the scope of this report and can be found elsewhere.^{20,21} Results of the history, review of systems, physical examination, and, in the case of ICH, neuroimaging are generally adequate to exclude these conditions. When there are concerns that a medical condition might be the cause of bruising or bleeding, the evaluation for the conditions in question should occur simultaneously with the evaluation for abuse.

Bruising

In the absence of independently witnessed accidental trauma or a known medical cause, any bruising in a nonmobile child is highly concerning for abuse and necessitates an evaluation for child abuse.^{12–15} Additionally, bruising in a young infant could also be the first presentation of a bleeding disorder.¹⁹ As such, a simultaneous evaluation for bleeding disorders is recommended in these cases. In mobile children, the locations and patterns of the bruising can be used to assess for the possibility of abuse (Table 1).

TABLE 1 Suspicion of Child Abuse in Ambulatory Children on the Basis of Characteristics of Bruises^{14,15,17}

Less Suspicious for Child Abuse	More Suspicious for Child Abuse
Forehead	Location
Under chin	Face
Elbows	Ears
Lower arms	Neck
Hips	Upper arms
Shins	Trunk
Ankles	Hands
	Genitalia
	Buttocks
	Anterior, medial thighs
	Pattern
	Slap or hand marks
	Object marks
	Bite marks
	Bruises in clusters
	Multiple bruises of uniform shape
	Large cumulative size of bruising

In cases of bruising, the assessment of the need for an evaluation for bleeding disorders should focus on the following:

- the specific history offered to explain the bruising;
- the nature and location of bruising; and
- mobility and developmental status of the child.

The following factors generally exclude the need for an evaluation for a bleeding disorder:

- the caregivers' description of trauma sufficiently explains the bruising;
- the child or an independent witness is able to provide a history of abuse or nonabusive trauma that explains the bruising; or
- abusive object or hand-patterned bruising is present.

The injury history offered by caregivers might be purposefully misleading if the caregivers have caused the bruising by abusive means.

In nonmobile infants, bleeding disorders can present with bruising or petechiae in sites of normal handling or pressure. Examples of this include the following:

- petechiae at clothing line pressure sites;
- bruising at sites of object pressure, such as in the pattern and location of infant seat fasteners; and
- excessive diffuse bleeding if the child has a severe bleeding disorder.

Absence of these examples does not rule out a bleeding disorder; however, their presence might increase the probability of a bleeding disorder.

ICH

Excepting obvious known trauma, ICH in a nonmobile child is highly concerning for child abuse. Children can

suffer ICH, such as a small subdural or an epidural hematoma underlying a site of impact, from a short fall; however, short falls rarely result in significant brain injury.¹⁶ Birth trauma and some medical conditions can also result in ICH in infants. Consultation with a child abuse pediatrician should be considered in complex or concerning cases.

No studies have systematically compared the presentation, clinical findings, patterns of ICH, or presence of retinal hemorrhages found in children with bleeding disorders with those found in children in whom abusive head trauma is diagnosed. However, bleeding disorders can cause ICH in any part of the cranial contents, and up to 12% of children and young adults with bleeding disorders have had ICH at some time.^{22,23} Children with ICH concerning for abuse require an evaluation for bleeding disorders. Exceptions to required evaluation can include the following:

- Independently witnessed or verifiable trauma (abusive or nonabusive),
- Other findings consistent with abuse, such as fractures, burns, or internal abdominal trauma.

Other Bleeding Symptoms

Children with conditions such as hematemesis, hematochezia, or oronasal bleeding as presenting symptoms should be evaluated on a case-by-case basis for possible abuse, particularly child abuse in a medical setting. Medical conditions and/or child abuse can cause these findings.

BLEEDING DISORDERS AND EXTENT OF EVALUATION

Bleeding disorders that can produce patterns of bruising or bleeding that mimic abuse include coagulation factor deficiencies/abnormalities, fibrinolytic

defects, defects of fibrinogen, and platelet disorders. Table 2 contains a listing of the most common bleeding disorders in children and characteristics of potential testing strategies for each disorder. Most factor deficiencies can be detected by the prothrombin time (PT) and activated partial thromboplastin time (aPTT); however, von Willebrand disease (VWD) and factor XIII deficiency are not reliably detected by these screening tests. Additionally, mild deficiencies in factor VIII or factor IX (mild hemophilia) might not cause abnormalities in the aPTT but might still result in significant bleeding, including ICH, particularly after mild trauma. Fibrinolytic defects can cause significant bleeding/bruising but are extremely rare and require specific testing. Defects of fibrinogen are also rare and can be detected by the fibrinogen concentration and thrombin time.

The prevalence of mild platelet disorders is unknown, and testing for mild platelet disorders is challenging. The most common clinical presentations include bruising and mucocutaneous bleeding. The prevalence of ICH in mild platelet disorders is unknown but is likely to be low. Platelet aggregation testing, best performed by a pediatric hematologist, requires a relatively large volume of blood, and interpretation of the test result requires a specialist.²⁵ A PFA-100 can screen for many platelet function disorders, including more severe types, such as Bernard Soulier syndrome and Glanzmann thrombasthenia, as well as many types of VWD. However, the PFA-100 is not an effective screen for some types of VWD and milder platelet abnormalities. Individual patient characteristics, such as hematocrit, platelet count, pregnancy, age, multisystem trauma, sepsis, and medications, can affect the results of the PFA-100. Accurate

diagnosis often requires additional testing, such as specific von Willebrand testing or platelet aggregation; therefore, many centers have decreased or ceased use of the PFA-100.^{25,26} Assessment of the results of a PFA-100 and the need for further testing are best accomplished in consultation with a pediatric hematologist.

Vitamin K Deficiency

Vitamin K deficiency in infants can result in bleeding in the skin or from mucosal surfaces from circumcision, generalized ecchymoses, large intramuscular hemorrhages, or ICH. Because of the widespread provision of vitamin K at birth, vitamin K deficiency bleeding (VKDB) is rare; however, not all states require vitamin K to be administered at birth, and some medical conditions predispose to VKDB.²⁴ In VKDB, there is a prolonged PT and possibly aPTT for age. In patients who have already received vitamin K, fresh-frozen plasma, or specific factor replacement as treatment, measurement of proteins induced by vitamin K absence can confirm the diagnosis.^{27,28}

Coagulation Tests in Cases of Bruising

The initial screening panel in a patient who presents with bruising evaluates for conditions with a known prevalence more common than 1 per 500 000 people, including idiopathic thrombocytopenic purpura, all factor deficiencies (except factor XIII deficiency), and VWD (Fig 1). It does not evaluate for extremely rare conditions, including factor XIII deficiency, defects of fibrinogen, and fibrinolytic defects. This strategy also does not screen for extremely rare platelet disorders, such as Glanzmann thrombasthenia, and more common but relatively more difficult to detect

platelet disorders, such as platelet storage pool disorders. If test results are abnormal or expanded/detailed testing is necessary or preferred, consultation with a pediatric hematologist is recommended.

In many circumstances, children with bruising that is suspicious for abuse may be removed from a potentially dangerous setting where the abuse likely occurred. A thorough physical examination performed in the weeks after removal that reveals minimal bruising and/or bruising only in locations of common accidental bruises is supportive of abuse as the cause of the original suspicious bruising. Each case must be evaluated individually, however, considering the totality of findings, and with the understanding that the need for safety must be balanced with the emotional trauma of removing a child from his or her home. Bleeding disorders are generally permanent conditions that do not result in abatement after a change in caregivers. One exception to this is immune thrombocytopenia (ITP), which is a transient, often self-resolving bleeding disorder. Screening for ITP (platelet count) is necessary at the time of presentation with bruises.

Determining the Need for a Test: The Medical Probability

Specific data regarding the prevalence of bleeding disorders in the population of children with ICH or subdural hematoma is not available. However, there are data regarding the probability of specific bleeding disorders to cause ICH. If the prevalence of a condition and the frequency of a particular presentation of that condition are known, a physician can construct the probability of that specific condition (bleeding disorder) resulting in the specific presentation (ICH). The presence of "classic" bleeding symptoms, such as bleeding after circumcision,

TABLE 2 Common Testing Strategies for Bleeding Disorders

Condition	Frequency	Inheritance	Screening Tests	Sn and Sp, %	PPV and NPV, %	Confirmatory Test
Factor abnormalities/deficiencies						
WVD type 1	1/1000	AD	PFA-100	Sn = 79–96 ^a	PPV = 93.3	WAg ^b WF activity WV multimer analysis Factor VIII activity
WVD type 2A	Uncommon	AD or AR	PFA-100	Sp = 88–96 ^a Sn = 94–100 ^a	NPV = 98.2 PPV = 93.3	WAg ^b WF activity WV multimer analysis Factor VIII activity
WVD type 2B	Uncommon	AD	PFA-100	Sp = 88–96 ^a Sn = 93–96 ^a	NPV = 98.2 PPV = 93.3	WAg ^b WF activity WV multimer analysis Factor VIII activity
WVD type 2M	Uncommon	AD or AR	PFA-100	Sp = 88–96 ^a Sn = 94–97 ^a	NPV = 98.2 PPV = 93.3	WAg ^b WF activity WV multimer analysis Factor VIII activity
WVD type 2N	Uncommon	AR, or compound heterozygote	aPTT	NA	NA	Factor VIII activity WF-Factor VIII binding assay
WVD type 3	1/300 000–1 000 000	AR, or compound heterozygote	PFA-100	Sn = 94–100 ^a	PPV = 93.3	WAg ^b Ristocetin cofactor WF multimer analysis Factor VIII activity
Factor II deficiency (prothrombin)	26 reported cases, estimated 1/1–2 million	AR, or compound heterozygote	aPTT, PT (may be normal)	Sn = variable	NA	Factor II activity ± antigen levels
Factor V deficiency	1/1 million	AR	aPTT, PT	Sn = variable	NA	Factor V activity
Combined Factor V/Factor VIII deficiency	1/1 million	AR	aPTT>PT	Sn = variable	NA	Factor V and factor VIII activities
Factor VII deficiency	1/300 000–500 000	AR	PT	Sn = variable	NA	Factor VII activity
Factor VIII deficiency	1/5000 male births	X-linked	aPTT	Sn = variable	NA	Factor VIII activity
Factor IX deficiency	1/20 000 male births	X-linked	aPTT	Sn = variable	NA	Factor IX activity
Factor X deficiency	1/1 million	AR	aPTT, PT, RVV	Sn = variable	NA	Factor X activity
Factor XI deficiency	1/100 000	AR	aPTT	Sn = variable	NA	Factor XI activity
Factor XIII deficiency	1/2–5 million	AR	Clot solubility	Sn = variable	NA	Factor XIII activity
Fibrinolytic defects						
α-2 antiplasmin deficiency	~40 reported cases	AR	Euglobin lysis test	Sn = variable	NA	α-2 antiplasmin activity
PAI-1 deficiency	Very rare	AR		Sn = variable	NA	PAI-1 antigen and activity
Defects of fibrinogen						
Afibrinogenemia	1/500 000	AR	PT, aPTT	Sn = high	NA	Fibrinogen level
Hypofibrinogenemia	Less than afibrinogenemia		PT, aPTT	Sn = variable	NA	Thrombin time, fibrinogen activity
Dysfibrinogenemia	1/million		Thrombin time, fibrinogen level	Sn = variable	NA	Thrombin time, fibrinogen antigen and activity level comparison, reptilase time
Platelet disorders						
ITP	Age-related	NA	CBC	Sn = high	NA	Antiplatelet Ab (rarely needed)
Glanzmann thrombasthenia	Very rare	AR	PFA-100	Sn = 97–100	NA	Platelet aggregation testing Flow cytometry

TABLE 2 Continued

Condition	Frequency	Inheritance	Screening Tests	Sn and Sp, %	PPV and NPV/%	Confirmatory Test
Bernard Soulier syndrome	Rare	AR	PFA-100	Sn = 100	NA	Platelet aggregation testing Flow cytometry
Platelet release/storage disorders	Unknown, more common than other platelet function disorders	variable	PFA-100	Sn = 27–50	NA	Platelet aggregation and secretion Electron microscopy Molecular and cytogenetic testing

AD, autosomal dominant; AR, autosomal recessive; CBC, complete blood cell (count); NA, not available or not applicable; NPV, negative predictive value; PAI-1, plasminogen activator inhibitor-1; PPV, positive predictive value; RV, Russell viper venom (test); Sn, sensitivity; Sp, specificity; VW, von Willebrand; VWAg, von Willebrand antigen; VWF, von Willebrand factor Ab, antibody.

^a Values derived from data before 2008 National Institutes of Health Consensus guidelines. Sn and Sp using current diagnostic cutoffs unknown but would be expected to have higher Sp with lower Sn.

^b May be reasonable to proceed directly to diagnostic testing depending on availability. See accompanying technical report for detailed discussion.²⁴

umbilical stump bleeding, joint hemorrhage, and excessive soft tissue bleeding, increase the probability for a bleeding disorder; however, these findings are neither sensitive nor specific for bleeding disorders.

Coagulation Tests in the Setting of ICH

For bleeding disorders that cause ICH, the prevalence of the bleeding disorder and the prevalence of ICH in patients with each specific bleeding disorder can be used to construct the probability of the specific bleeding disorder to cause ICH (Table 3). Some probabilities are so low as to preclude calculation. Testing for these conditions is likely not useful. Mild hemophilia, which might be missed if only an aPTT test is ordered, can be detected by measuring specific levels of factor VIII and factor IX. Mild hemophilia can result in ICH, particularly after mild trauma, and because of the relatively high prevalence of the condition, the probability of mild factor VIII deficiency causing or contributing to ICH is 1 in 280 000 males. In populations with a high prevalence of factor XI deficiency, such as the Ashkenazi Jewish population, it might be reasonable to measure factor XI level.

Clinical and historical information can be used to determine the need for testing in children with isolated ICH concerning for abuse (Fig 1). The initial testing panel for ICH evaluates for conditions for which the probability for the condition resulting in ICH is greater than 1 per 5 million. The panel includes testing for most factor deficiencies and afibrinogenemia. This screening panel does not test for factor XIII deficiency, VWD, fibrinolytic defects, hypofibrinogenemia, and dysfibrinogenemia. These conditions either have not been associated with ICH or they are so rarely the cause of ICH that testing for the conditions is

not reasonable. Additionally, the initial screening panel evaluates for disseminated intravascular coagulation (DIC). Because DIC can cause any type of bruising/bleeding, including ICH, the finding of DIC in the context of suspected child abuse could significantly change the clinical approach to a patient. In children with DIC and bleeding symptoms as the only finding concerning for abuse, consideration must be given to the multitude of primary causes of DIC, including trauma, sepsis, and primary bleeding disorders, among many others.

Many children with ICH suspicious for abuse, if they survive, are placed in safe settings after hospital discharge. In these cases, testing for bleeding disorders can be deferred to a later date, with the exception of ITP. If blood products have been given to the patient, as can happen in severe ICH, the definitive evaluation for bleeding disorders should be postponed until the transfused blood components are no longer in the patient's system (Table 4). Assistance from a pediatric hematologist should be considered in addressing the possibility of factor deficiencies after a transfusion has occurred.

Many aspects of bleeding disorders are under investigation, and thus, changes in the understanding of the prevalence and severity of certain bleeding symptoms related to these disorders should be expected. For example, although hemophilia A and B are X-linked diseases and, therefore, typically thought to affect only male individuals, 25% to 50% of female carriers of hemophilia report excess bleeding; therefore, measurement of factor VIII and IX levels in female patients should be considered.²⁹ In addition, the population prevalence and/or clinical effects of mild platelet function disorders continue to be studied. In a patient with mucocutaneous symptoms, particularly if petechiae are

TABLE 3 Probabilities for Congenital Coagulopathies Causing ICH^a

Condition	Prevalence of Condition, Upper Limits	Prevalence of ICH, Upper Limits	Probability ^b
VWD	1/1000	Extremely rare	Low
Factor II deficiency	1/1 million	11%	1/10 million
Factor V deficiency	1/1 million	8% of homozygotes	1/10 million homozygotes
Combined factors V and VIII deficiencies	1/1 million	2%	1/50 million
Factor VII deficiency	1/300 000	4%–6.5%	1/5 million
Factor VIII deficiency	1/5000 males	5%–12%	1/50 000 males
Factor IX deficiency	1/20 000 males	5%–12%	1/200 000 males
Factor X deficiency	1/1 million	21%	1/5 million
Factor XI deficiency	1/100 000	Extremely rare	Low
Factor XIII deficiency	1/2 million	33%	1/6 million
α-2 antiplasmin deficiency	40 cases reported	Not reported	Low
Plasminogen activator inhibitor-1 deficiency	Extremely rare	Common	Low
Afibrinogenemia	1/500 000	10%	1/5 million
Dysfibrinogenemia	1/1 million	Single case report	Low

^a The probability of having a specific bleeding disorder increases in the setting of a family history of that specific named bleeding disorder or if the patient is from an ethnicity in which a specific bleeding disorder is more common (eg, Ashkenazi Jewish people and factor XI deficiency).

^b "Probability" indicates the probability that an individual in the general population would have the following specific coagulopathy causing an ICH.

present, platelet aggregation testing should be considered.²⁵ Finally, because von Willebrand factor is an acute phase reactant, its levels can vary in response to clinical status, resulting in falsely elevated results. Many times, testing must be repeated up to 3 times to ensure reliable results.³⁰ If significant concern for VWD exists, consultation with a pediatric hematologist is suggested.

When Testing Indicates a Possible Bleeding Disorder in the Context of an Abuse Evaluation

Positive laboratory test results require further evaluation for the possibility of false-positive results and/or the ne-

cessity for further testing. Prolongation of the PT and aPTT because of parenchymal damage has been noted in abusive head trauma and should not automatically be interpreted as evidence of a primary bleeding disorder.³¹ Additionally, consideration must be given to the likelihood of a preexisting bleeding disorder as the primary cause of a child's bleeding/bruising. For example, given the relatively high prevalence of VWD, it is inevitable that some children with VWD will be abused and present with bleeding/bruising symptoms. Determining the causative factor in these situations is challenging. Bruising is a common finding in VWD. If a child has test results consistent with VWD and bruising concerning for abuse, a short-term change in home setting may be considered, understanding the cautions needed when using this approach. Only a few case reports have attributed ICH to VWD. Most reported ICH in children with VWD would not be confused with typical abusive ICH.^{32–34} Given the rarity of ICH in VWD, particularly spontaneous ICH, testing consistent with VWD does not mean that ICH is definitively attributable to VWD, and abuse must still be considered.

Interpretation of Tests

It should be noted that the aPTT can be falsely prolonged in certain circumstances, such as in the presence of a lupus anticoagulant, or can be prolonged and might not indicate a true bleeding disorder, such as in factor XII deficiency or other contact factor deficiencies. In addition, patients who experience a traumatic brain injury often have a transient coagulopathy that does not reflect an underlying congenital disorder.^{31,35} Coagulation tests are very sensitive to specimen handling and should be performed in laboratories experienced with these assays. Inappropriate handling commonly leads to false-positive results.

CONCLUSIONS

Children who present with bleeding and bruising symptoms that are concerning for abuse require careful evaluation for the potential of bleeding disorders as a cause. No single panel of tests rules out every possible bleeding disorder. Given the rarity of most bleeding disorders and the possible presence of specific clinical factors that decrease the likelihood of a bleeding disorder causing a child's findings, in many situations, extensive laboratory evaluation is not

TABLE 4 Half-Lives of Coagulation Factors

Factor	Half-Life Postinfusion, h
Fibrinogen	96–150
II	60
V	24
VII	4–6
VIII	11–12
IX	22
X	35
XI	60
XIII	144–300
VWF	8–12

VWF, von Willebrand factor.

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necessary. If a laboratory evaluation is conducted, tests should be chosen on the basis of the prevalence of the condition, patient and family history, ease of testing, blood volume required for testing, and, in the case of ICH, probability of a bleeding disorder causing ICH. Further consultation with a pediatric hematologist is recommended if specific, expanded testing is necessary, if preliminary testing suggests the presence of a bleeding disorder, if testing to rule out a specific bleeding disorder is needed, or if testing for very rare conditions is preferred.

GUIDANCE FOR PEDIATRICIANS

In children who have bruising or bleeding that is suspicious for abuse,

1. Complete medical, trauma, and family histories and a thorough physical examination are critical tools in evaluating for the possibility of abuse or medical conditions that predispose to bleeding/bruising.
2. In each case, careful consideration of the possibility of a medical condition causing the bleeding/bruising is essential. Specific elements of the history and characteristics of the bleeding/bruising can be used to determine the need for a laboratory evaluation for bleeding disorders.
3. If the evaluation indicates a need for laboratory testing for bleeding disorders, initial testing is focused

on the prevalence of the condition and potential of each specific condition to cause the specific findings in a given child (Fig 1).

4. Laboratory testing suggestive or indicating the presence of a bleeding disorder does not eliminate abuse from consideration. In children with bruising and laboratory testing suggestive of a bleeding disorder, a follow-up evaluation after a change in home setting can provide valuable information regarding the likelihood of a bleeding disorder causing the concerning findings.
5. Children with ICH often receive blood product transfusions. It is suggested that screening for bleeding disorders in these patients be delayed until elimination of the transfused blood clotting elements.
6. The discovery of new information regarding condition prevalence, laboratory testing, and clinical presentations of bleeding disorders is to be expected. Close collaboration with a pediatric hematologist is necessary to ensure the most current evaluation and testing methods.

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REFERENCES

1. Lieder HS, Irving SY, Mauricio R, Graf JM. Munchausen syndrome by proxy: a case report. *AACN Clin Issues*. 2005;16(2):178–184
2. Ulinski T, Lhopital C, Cloppet H, et al. Munchausen syndrome by proxy with massive proteinuria and gastrointestinal hemorrhage. *Pediatr Nephrol*. 2004;19(7):798–800
3. Stricker T, Lips U, Sennhauser FH. Oral bleeding: Child abuse alert. *J Paediatr Child Health*. 2002;38(5):528–529
4. Walton LJ, Davies FC. Nasal bleeding and non-accidental injury in an infant. *Arch Dis Child*. 2010;95(1):53–54
5. Paranjothy S, Fone D, Mann M, et al. The incidence and aetiology of epistaxis in infants: a population-based study. *Arch Dis Child*. 2009;94(6):421–424
6. McIntosh N, Mok JY, Margerison A. Epidemiology of oronasal hemorrhage in the first 2 years of life: implications for child

- protection. *Pediatrics*. 2007;120(5):1074–1078
7. Goodnight S, Hathaway W. *Disorders of Hemostasis and Thrombosis: A Clinical Guide*. 2nd ed. New York, NY: McGraw-Hill; 2001
 8. Anderst JD, Kellogg N, Jung I. Is the diagnosis of physical abuse changed when Child Protective Services consults a Child Abuse Pediatrics subspecialty group as a second opinion? *Child Abuse Negl*. 2009; 33(8):481–489
 9. O'Hare AE, Eden OB. Bleeding disorders and non-accidental injury. *Arch Dis Child*. 1984; 59(9):860–864
 10. Scimeca PG, Cooper LB, Sahdev I. Suspicion of child abuse complicating the diagnosis of bleeding disorders. *Pediatr Hematol Oncol*. 1996;13(2):179–182
 11. Jenny C, Hymel KP, Ritzen A, Reinert SE, Hay TC. Analysis of missed cases of abusive head trauma. *JAMA*. 1999;281(7):621–626
 12. Sugar NF, Taylor JA, Feldman KW; Puget Sound Pediatric Research Network. Bruises in infants and toddlers: those who don't bruise rarely bruise. *Arch Pediatr Adolesc Med*. 1999;153(4):399–403
 13. Carpenter RF. The prevalence and distribution of bruising in babies. *Arch Dis Child*. 1999;80(4):363–366
 14. Maguire S, Mann MK, Sibert J, Kemp A. Are there patterns of bruising in childhood which are diagnostic or suggestive of abuse? A systematic review. *Arch Dis Child*. 2005;90(2):182–186
 15. Jenny C, Reese R. Cutaneous manifestations of child abuse. In: Reese RM, Christian CW, eds. *Child Abuse Medical Diagnosis and Management*. 3rd ed. Chicago, IL: American Academy of Pediatrics; 2009:19–51
 16. Chadwick DL, Bertocci G, Castillo E, et al. Annual risk of death resulting from short falls among young children: less than 1 in 1 million. *Pediatrics*. 2008;121(6):1213–1224
 17. Dunstan FD, Guildea ZE, Kontos K, Kemp AM, Sibert JR. A scoring system for bruise patterns: a tool for identifying abuse. *Arch Dis Child*. 2002;86(5):330–333
 18. Feldman KW. Patterned abusive bruises of the buttocks and the pinnae. *Pediatrics*. 1992;90(4):633–636
 19. Jackson J, Carpenter SL, Anderst JD. Challenges in the evaluation for possible abuse: presentations of congenital bleeding disorders in childhood. *Child Abuse Negl*. 2012;36(2):127–134
 20. Sirotnak AP. Medical disorders that mimic abusive head trauma. In: Frasier L, Rauth-Farley K, Alexander R, Parrish R, eds. *Abusive Head Trauma in Infants and Children*. St Louis, MO: G W Medical Publishing; 2006: 191–214
 21. Dinehart SM, Henry L. Dietary supplements: altered coagulation and effects on bruising. *Dermatol Surg*. 2005;31(7 pt 2):819–826, discussion 826
 22. Mishra P, Naithani R, Dolai T, et al. Intracranial haemorrhage in patients with congenital haemostatic defects. *Haemophilia*. 2008;14(5):952–955
 23. Nelson MD, Jr, Maeder MA, Usner D, et al. Prevalence and incidence of intracranial haemorrhage in a population of children with haemophilia. The Hemophilia Growth and Development Study. *Haemophilia*. 1999; 5(5):306–312
 24. Carpenter SL, Abshire T, Anderst JD; American Academy of Pediatrics, Section on Hematology/Oncology and Committee on Child Abuse and Neglect. Technical report: evaluation for conditions that predispose to bleeding when child abuse is suspected. *Pediatrics*. 2012; (in press)
 25. Hayward CPM, Rao AK, Cattaneo M. Congenital platelet disorders: overview of their mechanisms, diagnostic evaluation and treatment. *Haemophilia*. 2006;12(suppl 3): 128–136
 26. Nichols WL, Hultin MB, James AH, et al. von Willebrand disease (VWD): evidence-based diagnosis and management guidelines, the National Heart, Lung, and Blood Institute (NHLBI) Expert Panel report (USA). *Haemophilia*. 2008;14(2):171–232
 27. Shearer MJ. Vitamin K deficiency bleeding (VKDB) in early infancy. *Blood Rev*. 2009;23 (2):49–59
 28. Miyasaka M, Nosaka S, Sakai H, et al. Vitamin K deficiency bleeding with intracranial hemorrhage: focus on secondary form. *Emerg Radiol*. 2007;14(5):323–329
 29. Plug I, Mauser-Bunschoten EP, Bröcker-Vriends AH, et al. Bleeding in carriers of hemophilia. *Blood*. 2006;108(1):52–56
 30. National Heart, Lung, and Blood Institute. *The Diagnosis, Evaluation and Management of von Willebrand Disease*. Bethesda, MD: National Heart, Lung, and Blood Institute, National Institutes of Health, US Department of Health and Human Services; December 2007. NIH Publication No. 08-5832
 31. Hymel KP, Abshire TC, Luckey DW, Jenny C. Coagulopathy in pediatric abusive head trauma. *Pediatrics*. 1997;99(3):371–375
 32. Ragni MV, Bontempo FA, Hassett AC. von Willebrand disease and bleeding in women. *Haemophilia*. 1999;5(5):313–317
 33. Ziv O, Ragni MV. Bleeding manifestations in males with von Willebrand disease. *Haemophilia*. 2004;10(2):162–168
 34. Mizoi K, Onuma T, Mori K. Intracranial hemorrhage secondary to von Willebrand's disease and trauma. *Surg Neurol*. 1984;22 (5):495–498
 35. Talving P, Lustenberger T, Lam L, et al. Coagulopathy after isolated severe traumatic brain injury in children. *J Trauma*. 2011;71(5):1205–1210

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