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Bleeding is a common chief complaint in the emergency department.

Recognizing excessive bleeding and treating bleeding disorders may be challenging. Awareness of the differential, diagnostic strategies and treatment options is critical. This article presents a comprehensive approach to the patient with a bleeding disorder.

— The Editor

Introduction

Appropriate hemostasis requires a complex interaction between platelets, vascular surface factors and clotting factors. The initial phase of hemostasis involves vascular reaction of the injured blood vessel and the formation of a platelet plug (primary hemostasis). Clotting factors then form the fibrin clot that provides the permanent seal (secondary hemostasis). A defect in any

Clinical Presentation, Evaluation and Management of Bleeding Disorders in Children

Authors: **Beng R. Fuh, MD**, Assistant Professor of Pediatrics, Department of Pediatrics, Division of Pediatric Hematology and Oncology, Brody School of Medicine at East Carolina University; and **Ronald M. Perkin, MD**, Professor and Chairman, Department of Pediatrics, Brody School of Medicine at East Carolina University.

Peer Reviewer: **Afshin Ameri, MD**, Associate Professor of Pediatrics, Director of Pediatric Comprehensive Hemophilia Program, Medical College of Georgia, Augusta.

of the above factors can lead to abnormal bleeding. In addition, structural abnormalities such as hemangiomas or ectatic capillaries can lead to excessive bleeding. Bleeding disorders vary widely in severity and it can be challenging to establish which bleeding is excessive and which is within acceptable limits. It is not unusual for patients with congenital bleeding disorders to remain undiagnosed into their adulthood. Patients with excessive bleeding must be assessed for possible bleeding disorders and targeted treatment provided to prevent or minimize serious complications. Assessment should include a careful clinical history and laboratory evaluation. Disorders of primary hemostasis frequently present with epistaxis, bleeding of mucous membranes and bruising. Disorders of secondary hemostasis frequently present with soft

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tissue bleeds, hemarthrosis and large vessel bleeding. Management strategies vary from observation, local control, infusions of factor products to physical therapy and rehabilitation following bleeding complications.

Epidemiology and Etiology

Excessive bleeding secondary to primary defects in the coagulation system is rare and usually due to congenital quantitative or qualitative deficiencies in one of the components of the coagulation system. Patients can develop inhibitors following infections or medication leading to transient acquired bleeding disorders. The majority of cases of excessive bleeding in children are due to secondary causes such as vascular abnormalities or medication effects such as aspirin, ibuprofen, clopidogrel, etc. Inappropriate activation of the coagulation cascade can lead to platelet and coagulation factor consumption such as occurs in disseminated intravascular coagulation (DIC).

von Willebrand Disease (vWD) is the most common inherited bleeding disorder affecting about one to two percent of Americans.^{1,2} It is usually inherited in an autosomal dominant mode though some variants are autosomal recessive. von Willebrand antigen activates platelets by binding to the GP1b region of platelets. It also binds FVIII thereby stabilizing it. Very low levels of von Willebrand antigen can lead to low FVIII levels and inadequate formation of the platelet plug. vWD type IIB

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Please call **Allison Weaver**,
Managing Editor, (617) 629-5951, or e-mail
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Table 1. Causes of Thrombocytopenia

DISORDERS OF DECREASED PRODUCTION	DISORDERS OF INCREASED DESTRUCTION	OTHERS
• Bone marrow failure syndromes	• ITP	• Hypersplenism
• Hereditary	• DIC	• Drug-induced
• Marrow infiltration	• vWD type IIB	• Gestational
• Chemotherapy-induced	• Heparin-induced	• Infection-related
• Radiation therapy-induced	• TTP/HUS	• Hemophagocytosis
	• NAIT	
	• Mechanical destruction	

Abbreviations: ITP—idiopathic thrombocytopenic purpura; DIC—disseminated intravascular coagulation; vWD—von Willebrand Disease; TTP—thrombotic thrombocytopenic purpura; HUS—hemolytic uremic syndrome; NAIT—neonatal alloimmune thrombocytopenia

results in enhanced binding affinity of von Willebrand antigen to platelets resulting in increased platelet clearance and thrombocytopenia.³ Other variants of vWD are due to defects in the binding site to FVIII or absence of certain multimeric fractions. vWD can also arise from the development of autoimmune antibodies to the von Willebrand factor. Conditions associated with von Willebrand factor auto antibody formation include leukemia, SLE, and lymphoma.⁴ Wilms tumor⁵ and hypothyroidism⁶ have been associated with decreased synthesis of von Willebrand factor.

Hemophilia A (FVIII deficiency) has an incidence of 1 in 5,000 males. The gene for factor VIII is located on the X chromosome. Female carriers are usually unaffected, though extreme lyonization of the unaffected X chromosome, Turner's Syndrome, or daughters of affected fathers and carrier mothers can rarely lead to symptomatic females. Lyonization is the process whereby one X chromosome in each cell is randomly inactivated. It is described as severe when FVIII activity level is 1%, moderate if FVIII activity level is between 1% and 5%, and mild if FVIII is 5% to 40%.

Hemophilia B (FIX deficiency) is also X-linked and has an incidence of about 1 in 40,000 males.

Both hemophilia A and B have no racial predilection, and about 30% of patients are due to new mutation with no prior family history of hemophilia.⁷

FXI deficiency, also referred to as hemophilia C, is inherited as an autosomal recessive condition. It is a very rare condition mostly found in people of Jewish ancestry.⁸

Other specific factor deficiencies include FXIII deficiency, FVII deficiency, hypofibrinogenemia, dysfibrinogenemias, etc.

Vitamin K deficiency leads to a decrease in vitamin-K-dependent coagulation factors and can lead to bleeding. Maternal use of anti-convulsants are a common cause of early-onset hemorrhagic disease of the newborn. Late-onset hemorrhagic disease of the newborn usually occurs in exclusively breastfed



Table 2. Drugs Associated with Thrombocytopenia

ANTIPLATELET AGENTS	ANTIMICROBIALS	CARDIOVASCULAR AGENTS	NEUROPSYCHIATRIC AGENTS	OTHER
Anagrelide	Amphotericin	Amiodarone	Carbamazepine	Gold
Abciximab	Ampicillin	Captopril	Chlorpromazine	Heparin
Eptifibatide	Isoniacid	Digoxin	Diazepam	
Ticlopidine	Rifampin	Hydrochlorothiazide	Haldoperidol	
Tirofiban	Methicillin	Procainamide	Lithium	
Acetaminophen	Piperacillin	Quinidine	Methyldopa	
Diclofenac	Sulfisoxazole		Phenytoin	
	Bactrim			

Note: This list is not exclusive and includes only common drugs. When confronted with thrombocytopenia of unclear etiology, it is important to always review the patient's medications for possible effects on platelet count.

Adapted from Deloughery T. Hemorrhagic and thrombotic disorders in the intensive care setting. In: Kitchens C, Alving BM, et al, (eds). *Consultative Hemostasis and Thrombosis*. Philadelphia: W.B. Saunders Co.;2002:493-513.

infants with inadequate neonatal vitamin K prophylaxis. In older children, vitamin K deficiency can result from malabsorption syndromes, prolonged emesis, and warfarin use.

Liver disease causes a decrease in the synthesis of clotting factors, the most sensitive of which are FV and FVII.

Thrombocytopenia secondary to congenital amegakaryocytic thrombocytopenia, thrombocytopenia absent radius (TAR), etc. are rare. Fanconi anemia and other forms of aplastic anemia are other rare causes of thrombocytopenia. Peripheral consumption or destruction or bone marrow infiltration such as occurs in leukemia can also lead to thrombocytopenia. **Table 1** shows the common causes of thrombocytopenia, and **Table 2** shows drugs frequently associated with thrombocytopenia.⁹

The Coagulation Cascade

Most coagulation factors are synthesized in the liver. Factors II, VII, IX, X, and XII are synthesized as inactive enzymes that become active when cleaved. Factors II, VII, IX, and X are vitamin K dependent. Activation of coagulation factors culminates in the generation of thrombin, which in turn cleaves fibrinogen to produce fibrin and activates platelets. **Figure 1** shows a scheme of the coagulation cascade.

Clinical Presentation

Clinical presentation varies depending on the etiology of the bleeding disorder. It can be difficult to differentiate between normal bleeding and excessive bleeding. A very detailed bleeding history, including a family history, is critical in the diagnosis of a bleeding disorder. If there is a family history of excessive bleeding, a pedigree should be developed to identify a possible hereditary disease. Disorders of primary hemostasis usually present with epistaxis, mucosal membrane bleeding, and superficial ecchymosis, while disorders of secondary hemostasis usually present with joint bleeding, intramuscular bleeding, or other large-vessel bleeding.

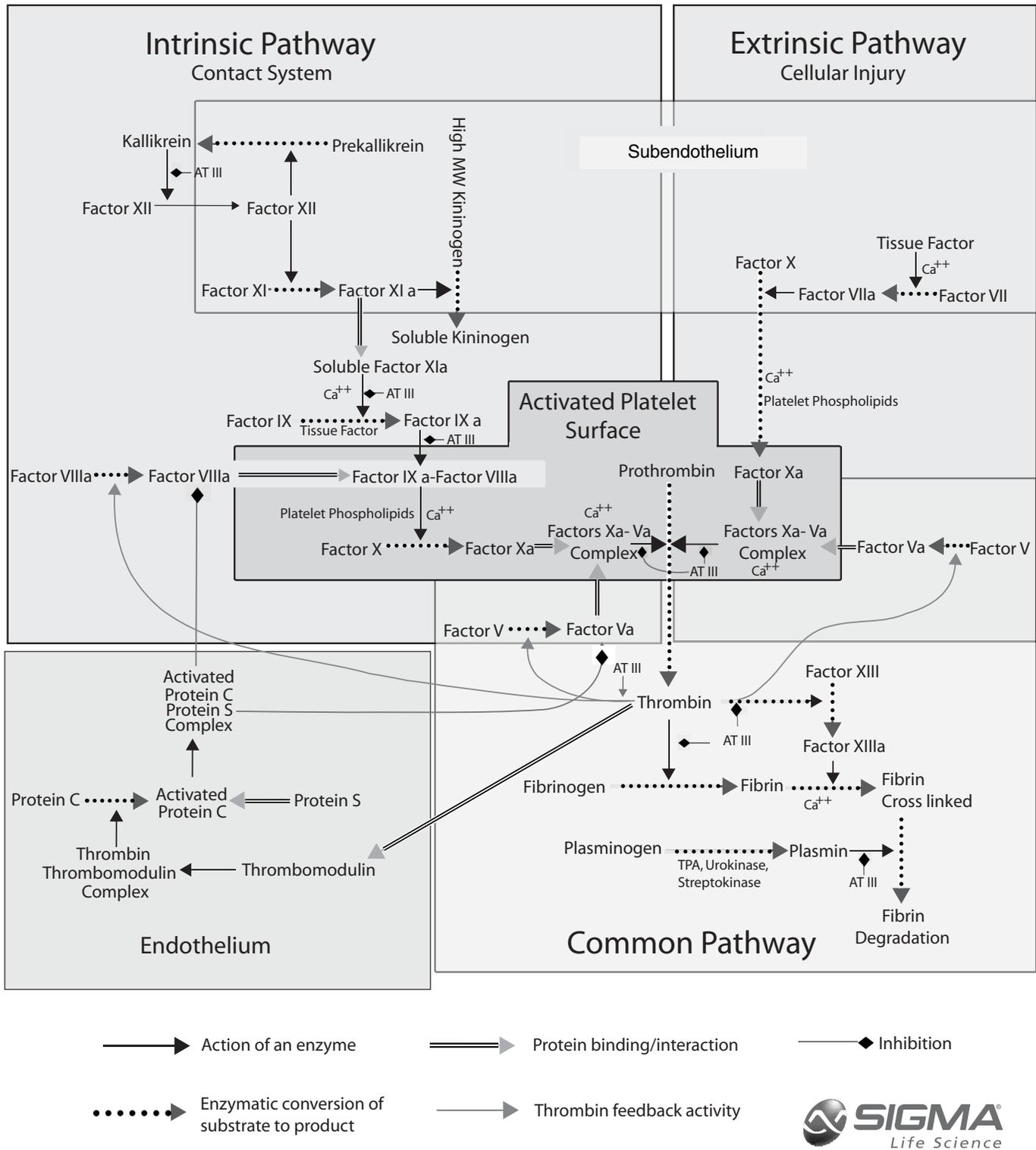
Thrombocytopenia. Thrombocytopenia usually manifests itself as petechiae, ecchymoses, and mucous membrane bleeding. There can also be excessive immediate bleeding following surgery or trauma. Bleeding is rare at platelet counts greater than 50,000/mm³, in the absence of a platelet dysfunction. The risk of bleeding is also higher in disorders of decreased production, such as chemotherapy-induced thrombocytopenia, than in disorders of increased platelet destruction such as ITP. The PT and aPTT are usually normal.

von Willebrand Disease. Most patients with vWD are asymptomatic. When symptomatic, it is usually as mucocutaneous bleeding such as menorrhagia in females, epistaxis, or ecchymosis. Occasionally, patients may present with excessive post-surgical bleeding, such as post tonsillectomy. Type III vWD may present with symptoms similar to hemophilia A or B. Since most forms of vWD are inherited in an autosomal dominant fashion, a good family history often reveals other family members with symptoms of excessive bleeding. APTT is usually prolonged, but may be normal, especially in mild vWD. von Willebrand factor (vWF) is an acute phase reactant, and even the stress of impending phlebotomy may be enough to increase vWF levels.¹⁰

Hemophilia A. Patients can present with a wide range of bleeding manifestations, from easy bruising to joint bleeds to intracranial bleeding. Recurrent joint bleeding can lead to target joints with the risk of permanent injury. In the neonatal period, hemophilia A can present as excessive post-circumcision bleeding, excessive bleeding post-venipuncture, or intracerebral hemorrhage. Affected individuals are usually male, and there is usually history of affected males in the maternal family. Rarely, females may be affected secondary to extreme lyonization, Turner's Syndrome, daughter of a carrier mother, or daughter of a hemophiliac father and carrier mother.

Hemophilia B. Hemophilia B has a similar presentation as hemophilia A. **Table 3** summarizes the common complications of hemophilia based on severity.

Figure 1. Scheme of the Coagulation Cascade



The extrinsic and intrinsic pathways should not be understood as two separate pathways, but are interdependent pathways which interact to produce a more efficient coagulation. (Roman numerals denote clotting factors.)
Used with permission from Sigma-Aldrich, St. Louis, MO.

Table 3. Characteristics of Hemophilia Bleeding Based on Severity

Aspect	Mild (5%–40%)	Moderate (2%–5%)	Severe (1%)
Age of onset	Usually after 3 yrs	Usually before 2 yrs	Usually first yr
Musculoskeletal bleeding	Joint/muscle bleeding rare, except in major trauma	Joint/muscle bleeding with minor trauma; “target joints” rare	Spontaneous joint/muscle bleeding; “target joints” frequent
Postoperative bleeding	Asymptomatic, but wound oozing or hematomas may occur	Wound hematomas common	Brisk bleeding common
Neonatal complications	Usually none	Post-circumcision bleeding may occur; ICH is rare	ICH, post-circumcision bleeding may occur
ICH	Very rare, but may occur with significant trauma	May occur	Relatively common; occurs in 3% of pts
Bleeding with dental extraction	Frequently occurs	Commonly occurs	Almost always occurs
Development of inhibitors	Very rare; < 0.5% of pts	Rarely occurs; ~3% of pts	Very common; ~20% of pts
Response to DDAVP	Two- to three-fold level increase	~10 % increase in level	No response

Key: DDAVP – desmopressin; ICH – intracerebral hemorrhage; pts – patients; yr – year

A “target joint” is a joint that has experienced repeated bleeds and is at high risk of re-bleeding.

Adapted from: Dimichelle DM. Hemophilia A. In: Goodnight SH, Hathaway WE (eds), *Disorders of Hemostasis and Thrombosis, A Clinical Guide*. 2nd ed. New York: McGraw Hill; 2001:1227–1139.

Evaluation

When confronted with acute or chronic bleeding, it is important to determine if the bleeding is within normal limits or is excessive. Evaluation should include a good history, including details on the locations, amount and frequency of bleeding, physical examination, and basic laboratory tests. Based on these findings, further testing may be done. Questions should be specific, and patients and parents should describe bleeding qualitatively and quantitatively, avoiding the terms “normal” or “abnormal.” Parents often base “normal” or “abnormal” on their own experiences; for example, a mother with menorrhagia secondary to undiagnosed vWD may describe excessive menstrual flow in her daughter as normal. **Table 5** is a suggested hemostatic history questionnaire.¹¹ Pictorial charts can be used to assist in assessing the amount of menstrual bleed.¹²

Physical examination can give very important clues as to the underlying disorder. Petechiae usually indicate

thrombocytopenia. Enlarged spleen and or enlarged liver may signify chronic illness or malignancy. Hyperextensibility of joints may indicate a connective tissue dysfunction, as in Ehlers-Danlos syndrome. Hemangiomas may suggest a consumptive etiology; joint abnormalities may suggest hemophilia.

Screening tests. If a bleeding abnormality is suspected, the following tests should be obtained. Attention must be paid to proper collection of samples including the right plasma to reagent volume and the avoidance of heparin contamination. Reference values are age dependent and vary from laboratory to laboratory which is a function of reagents used. It is important to have this in mind when interpreting results. For example aPTT is usually prolonged in neonates. Most bleeding factors reach adult levels by age 6–12 months.

Complete blood count (CBC). This will determine if there is a quantitative platelet disorder and if only platelets are affected. In bone marrow failure syndromes or infiltrative marrow processes, there are often abnormalities in other hematopoietic cell lines. The size of platelets should also be assessed. Large

Factor XIII Deficiency. Factor XIII deficiency may present with bleeding soon after birth, or as delayed post-surgical bleeding; for example, 24–72 hours post-tonsillectomy.

Factor XI and XII. Factor XI and XII deficiencies are usually asymptomatic, or there may be mild bleeding.

Disorder of fibrinogen. Disorders of fibrinogen may manifest in different ways depending on severity, as shown in **Table 4**.

Vitamin K deficiency. Vitamin K deficiency can present as early- or late-onset hemorrhagic disease of the newborn, with intracerebral hemorrhage, cephalohematoma, etc. Older children can present with mild to severe bleeding; PT and aPTT are usually prolonged.

Disseminated Intravascular Coagulation (DIC). Clinical symptoms of DIC may be masked by the underlying disease, making a high level of suspicion and evaluation necessary. Bleeding diathesis is usually more pronounced than microthrombosis. Oozing from intravenous catheter sites, ecchymosis, and petechiae are frequent manifestations.

Table 4. Clinical Manifestation of Disorders of Fibrogen Based on Severity

SEVERITY	CLINICAL MANIFESTATIONS
Afibrinogenemia	Severe, lifelong hemorrhagic diathesis
Hypofibrinogenemia	Mild bleeding—menorrhagia, post-operative bleeding, etc.
Dysfibrinogenemia	Ranges from asymptomatic to mild bleeding to thrombosis

Table 5. A Hemostasis Questionnaire: History to Obtain from Patient or Parent

1. *If abnormal bruising (ecchymosis or petechiae):* Are bruises extensive (larger than a quarter, indurated), located where trauma is unlikely, and unexplained by minor injury? Are petechiae ever seen? Is there gum bleeding?
2. *If prolonged bleeding after laceration or surgery:* Has there been prolonged (hours) or recurrent bleeding after lacerations (cuts, oral injury), surgery (circumcision, skin biopsy, tonsillectomy), tooth extractions? Poor wound healing? List all operations and significant trauma.
3. *If epistaxis or menorrhagia:* Has there been prolonged and heavy menstrual bleeding or severe recurrent epistaxis? If so, was anemia present or need for iron therapy or transfusion?
4. *If soft tissue or joint hemorrhage:* Is there a history of unusual hematomas or unexplained arthritis or joint swelling?
5. Has there been hematemesis, melana, hematuria, or hemoptysis without obvious cause?
6. *Family history.* Has any blood relative had a problem with excessive bleeding as noted in the questions above?
7. *General health:* Is there evidence of a disorder known to be associated with a bleeding tendency (e.g., liver disease, renal disease, connective tissue disorder, leukemia, SLE, malabsorption syndrome, myeloproliferative disorder, amyloidosis)? Is there evidence for abuse or self-inflicted injury?
8. *Drugs or medication:* Has aspirin, an antibiotic, or warfarin been taken in the last 10–14 days? Has vitamin K been used? History of transfusion?

Adapted from Goodnight SH, Hathaway WE (eds). Evaluation of bleeding tendency in the outpatient child and adult. In: *Disorders of Hemostasis and Thrombosis, A Clinical Guide*. 2nd ed. New York: McGraw Hill;2001: 52-60.

platelets often denote an active bone marrow as seen in processes associated with peripheral platelet destruction such as occurs in ITP. Normal platelets are seen in marrow suppression or failure and small platelets may be seen in Wiskott Aldrich Syndrome (WAS).

Prothrombin Time (PT)/International Normalized Ratio (INR). This evaluates the extrinsic pathway of the coagulation

Table 6. Common Bleeding Disorders and Basic Laboratory Findings

DISORDER	PLATELET COUNT	PT	APTT
Thrombocytopenia	Low	Normal	Normal
Platelet dysfunction	Normal	Normal	Normal
Hemophilia	Normal	Normal	Prolonged
FVII deficiency	Normal	Prolonged	Normal
vWD	Usually normal	Normal	Normal to prolonged
Vitamin K deficiency	Normal	Prolonged	Prolonged
Liver disease	Low to normal	Prolonged	Prolonged
DIC	Low	Prolonged	Prolonged
Vascular anomaly	Normal	Normal	Normal

Key: DIC – disseminated intravascular coagulation; PT – prothrombin time; aPTT – activated partial thromboplastin time

cascade. Factor VII is unique to the extrinsic pathway so a factor VII deficiency would result in a prolonged PT and normal aPTT. INR normalizes the variability in the sensitivities of thromboplastin reagents to low concentrations of some coagulation proteins. It is normally used to monitor oral anticoagulant therapy.

Activated Partial Thromboplastin Time (aPTT). Assesses the intrinsic pathway of the coagulation cascade. The aPTT is usually normal until factors levels drop to less than 30%.

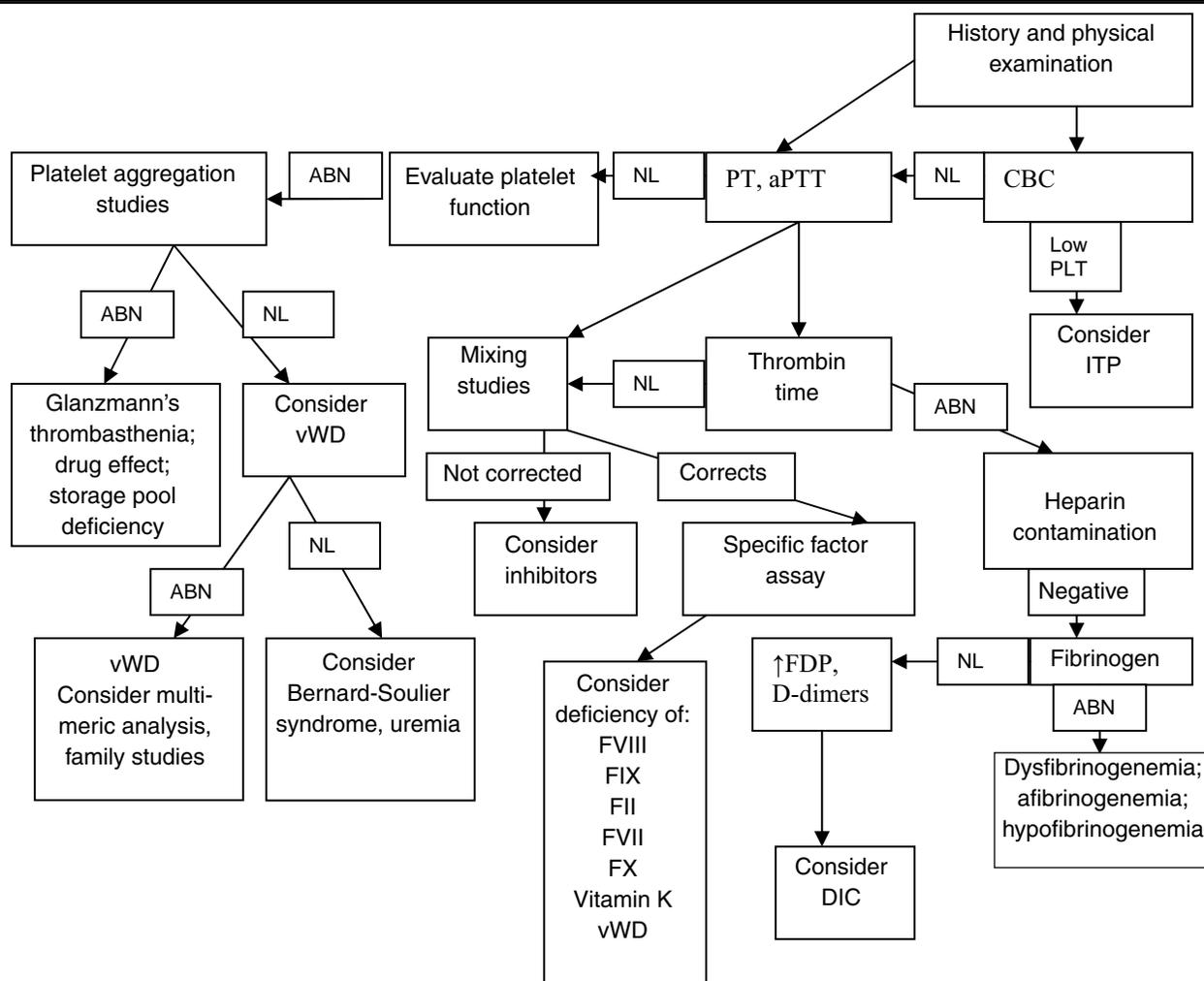
Table 6 shows the common differential diagnoses based on platelet count, PT, and aPTT findings.

If the above findings suggest a disorder of hemostasis, specific testing should be done. **Figure 2** shows a suggested decision-making tree for the evaluation of a hemostatic disorder.¹³

Idiopathic thrombocytopenic purpura (ITP). If a CBC shows isolated thrombocytopenia, ITP is very likely, especially if the smear reveals large platelets. The patient should be managed presumptively as ITP, and no further testing is necessary.

von Willebrand disease. von Willebrand antigen and activity levels can fluctuate and repeat testing is frequently necessary. Individuals with blood group O generally have lower vWF levels. A discrepancy between von Willebrand antigen level and activity level (ristocetin cofactor) may suggest a qualitative vWD subtype. In this case, multimeric analysis and platelet aggregation studies are important. Care should also be taken when interpreting von Willebrand panel results in patients who are on birth control, as this may artificially increase the level of von Willebrand antigen level. von Willebrand antigen level is a quantitation of vWF using immunoelectrophoresis or ELISA.¹⁴ Ristocetin induces the binding of vWF to the glycoprotein Ib receptor of platelets; this is used to measure the activity of vWF. vWF carries FVIII in circulation, thereby protecting it from degradation, and its level is helpful in diagnosing vWD. vWF is composed of different multimeric sizes, and these can be separated via gel electrophoresis. The distribution is normal in type I vWD, while type IIA vWD has a decrease in large multimers.

Figure 2. Suggested Decision Tree for the Evaluation of a Child with a Bleeding Disorder



Note: Bold captions denote findings. Arrows point to recommended actions or possible diagnosis.

Key: PT—prothrombin time; aPTT—activated partial thromboplastin time; F—factor; CBC—complete blood count; PLT—platelet count (normal white blood count and hemoglobin are assumed); vWD—von Willebrand disease; FDP—fibrin degradation products; DIC—disseminated intravascular coagulation; NL—normal; ABN—abnormal; ITP—idiopathic thrombocytopenic purpura

Adapted from Hastings CA, Lubin BH. Blood. In: Rudolph AM, Kamei RK(eds), *Rudolph's Fundamentals of Pediatrics*, 2nd ed. Norwalk, CT: Appleton & Lange. 1998: 441-490

Management of Bleeding Disorders

In cases of active bleeding priority has to be controlling the bleeding and maintaining the airway and circulation. Where possible, compression and pressure should be applied, with caution taken to limit damage to vital structures. If the patient has a known specific hemostatic defect, specific treatment should be administered as soon as possible. Generally patients with established hemophilia should carry an alert bracelet bearing their diagnosis. If the patient is hemodynamically unstable, consider transfusion with packed red blood cells (PRBC), platelets, and fresh frozen plasma (FFP).

Hemophilia. Patients with known severe hemophilia need to

be treated promptly whenever CNS bleeding is suspected or possible — for example, after head trauma — even if there is no obvious sign of bleeding. Treatment should be administered prior to imaging studies.

Patients who develop bleeding secondary to other conditions need treatment for the primary condition in addition to managing active bleeding. Avoid intramuscular (IM) injections. In patients with mild hemophilia A, Desmopressin (DDAVP) 0.3 mcg/kg may be used if the patient is known to be a responder. Desmopressin causes the release of vWF and FVIII from storage sites in endothelial cells, thereby raising serum FVIII levels. DDAVP can raise FVIII levels by up to six-fold.¹⁴ However, it is difficult

Table 7. Dosing Guidelines for Hemophilia

TYPE OF BLEED	RECOMBINANT FVIII DOSE FOR HA	RECOMBINANT FIX DOSE FOR HB	OTHER RECOMMENDATIONS
Life-threatening (CNS, airway, hip, GI, retroperitoneal)	50 units/kg; treat suspected CNS bleed prior to imaging	100 units/kg; treat suspected CNS bleed prior to imaging.	CNS: If + CT, continuous factor infusion, 2–4 units/kg/hr for 10–14 days Eye: Consult ophthalmologist Airway: Maintain Compartment syndrome: Consider decompression
Iliopsoas	50 units/kg bolus, then 25 units/kg Q 12 for 10–14 days	80 units/kg bolus, then 40 units/kg QD for 10–14 days	Hospitalize; strict bed rest
Hemarthrosis	25 units/kg bolus; repeat dose in 24 hrs, then Q 48 hrs for 1 week	40 units/kg bolus, then Q 48 hrs for 1 week	RICE, radiographs, splinting
Mucocutaneous (mouth, tooth extraction, epistaxis, menorrhagia)	20 units/kg bolus; Amicar 100–200 mg/kg loading dose, then 50–100 mg/kg Q6 for 10 days	30 units/kg bolus; Amicar 100–200 mg/kg loading dose, then 50–100 mg/kg Q6 for 10 days	Epistaxis: Pressure for 15–20 minutes; consider nasal packing or cautery
Traumatic hematuria	50 unit/kg bolus; maintenance IVF; no Amicar	100 units/kg; maintenance IVF; no Amicar	Bed rest; consider continuous factor
Lacerations requiring sutures	20 units/kg bolus prior to sutures	30 units/kg prior to sutures	Ice and pressure
Pre-operative management depends on procedure and complications	50 units/kg 1 hr prior to surgery; consult hematologist for intra- and postoperative management	100 units/kg 1 prior to surgery; consult hematologist for intra- and postoperative management	Complete blood count, FVIII level, FIX level, and inhibitor titers

Key: RICE—rest, ice, compression, elevate; QD—every day; hr—hour; IVF—IV fluid; CNS—central nervous system; CT—computed tomography; GI—gastrointestinal

to predict serum levels of FVIII after the administration of DDAVP, especially in stress situations. DDAVP should therefore be used only before the patients arrives at the emergency department or when IV access cannot be established for factor concentrate administration. If repeat dosing is necessary, monitor factor VIII levels for response and monitor for tachyphylaxis and sodium levels. Avoid treating more often than every eight hours or using more than three doses per treatment course.¹⁴

Patients with moderate or severe hemophilia should be treated with FVIII or FIX products for hemophilia A and hemophilia B, respectively. **Table 7** shows suggested dosing guidelines for patients with factors VIII and IX deficiency for different types of bleeds. Wherever possible, recombinant products should be used because these have lower treatment risks.

Hemophilia patients with inhibitors. For patients with hemophilia A with inhibitors and non-life- or limb-threatening bleeding, high-dose FVIII (100 units/kg bolus followed by 20 units/kg/hr via continuous infusion should be administered). If bleeding persists, treat with activated FVII (Nvo 7, Novo

Nordisk). If bleeding is not controlled by FVII, consider prothrombin complex concentrates (PCC) at 75 units/kg.¹⁵ Monitor closely for DIC and thrombosis, and a hematologist should be consulted. Porcine FVIII is available for use in patients with inhibitors, but should only be used under the guidance of the hematologist.

For patients with hemophilia B with inhibitors, activated FVII at a dose of 100 mcg/kg (range 90–240mcg/kg) every 6 hours (range 2–12 hours) should be used. If bleeding persists, consider PCC at a dose of 75 units/kg and monitor closely for DIC and thrombosis.¹⁵ PCC should not be given to patients with a history of anaphylaxis to FIX. The hematologist should be consulted.

Important facts about factor FVIII and FIX replacement products. FVIII is available as recombinant products (recombinate, kogenate, helixate, advate, refacto, etc.), monoclonal purified products (hemofil-M, Monoclate, AHF-M), and porcine factor (Hyate-C). Whenever possible, recombinant factors should be used; it has a half life of 8–12 hours, and a 1 unit/kg dose

Table 8. Dosing Guidelines for vWD

TYPE OF BLEED	RISTOCETIN COFACTOR DOSE	OTHER RECOMMENDATIONS
Life-threatening (CNS, airway, hip, GI, retroperitoneal)	50 units/kg; treat suspected CNS bleed prior to imaging	CNS: If + CT, continuous infusion 3 units/kg/hr for 10–14 days Eye: Consult ophthalmologist Airway: Maintain Compartment syndrome: Consider decompression
Hemarthrosis, muscle, or significant subcutaneous	25 units/kg DDVP for responders	RICE, consider splinting
Lacerations	25 units/kg or DDAVP for responders prior to sutures	Compress to control bleeding; ice
Mucocutaneous (mouth, tooth extraction, epistaxis, menorrhagia)	25 units/kg or DDAVP for responders Amicar 100–200 mg/kg loading dose, then 50–100 mg/kg q 6 hrs for 10 days	Epistaxis: Pressure for 15–20 minutes; consider nasal packing or cauterly
Traumatic hematuria	50 units/kg, then consider continuous infusion at 2 units/kg/hr if severe	Bed rest; no Amicar; consider steroids
Pre-operative management depend on procedure and complications	50 units/kg 1 hr prior to surgery, or DDAVP for responders	Consult hematologist for intra- and postoperative management

Key: RICE—rest, ice, compression, elevate; QD—every day; hr—hour; DDAVP—desmopressin; IVF—IV fluid; CNS—central nervous system; CT—computed tomography; GI—gastrointestinal

increases plasma FVIII level by 2%. It is the treatment of choice in hemophilia A.

Recombinant FIX is available as recombinant products (benefix) or highly purified products (monomine, alphanine, etc.). It has a half life of 18–24 hours and a 1 unit/kg dose increases plasma FIX level by 0.75%–1%. It is used to treat hemophilia B.

Factor VII is available as a recombinant product. It has a half life of two hours and is used to treat FVII deficiency or hemophilia A or B patients with inhibitors.

Amicar (epsilon-aminocaproic acid) is an anti-fibrinolytic agent that helps stabilize the fibrin clot. Dosing is 100–200 mg/kg (max 10 g) loading dose followed by 50–100 mg/kg (max 5 g) every 6 hours, not to exceed 30 g/day. It is most useful for mucous membrane bleeding, especially nasal, oral, and gastrointestinal tract bleeding, where enzymes contained in secretions can accelerate clot degradation. It can be administered orally or parenterally and should be given every six hours for 7–10 days. For oral bleeding, it can be used as swish and spit. Swish for two minutes prior to expectoration. Patient should not eat or drink anything (remain NPO) at least one hour after swish and spit. Amicar is contraindicated in patients with hematuria.

Most patients with hemophilia receive comprehensive care at specialized regional centers, and studies have shown that this leads to significantly improved outcomes. Hemophilia treatment

centers generally have around-the-clock coverage to assist patients and parents, emergency department, and primary care physicians in case of emergency. Patients may carry a supply of their factor, and this can be used in case the treating physician does not have readily available factor products.

Von Willebrand Disease. *Desmopressin (DDAVP).* DDAVP acts by releasing vWF from storage sites in endothelial cells. This leads to a rise in serum levels of vWF and FVIII. Use 0.3 mcg/kg diluted in 25–50 mL normal saline infused over 30 minutes or nasal spray. DDAVP should be used with caution in patients with vWD type 2B, since it can worsen thrombocytopenia in this subset of patients. (See Table 8.)

Nasal DDAVP for vWD (Stimate) should not be confused with generic DDAVP used for enuresis or panhypopituitarism.

DDAVP dosing. For body weight < 50kg, give 1 puff DDAVP (150 mcg). For weight > 50kg, give 2 puffs (300 mcg). vWF has a half life of 8–12 hours.

Stimate is appropriate for home use and minor bleeding only. In the emergency department setting, cryoprecipitate or vWF-enriched plasma preparations should be used. Severe bleeding can also be treated with Humate-P or alphanate at 25 ristocetin cofactor units/kg.

Other treatment products. *FFP.* Use for disorders for which factor concentrates are not available. Dosing is 5–10 mL/kg; 1 mL of FFP ~ 1 unit of each clotting factor. For doses > 250 mL,

Table 9. Complications of Various Factor Replacement Products

COMPLICATION	PRODUCT			
	FFP or cryo-precipitate	Intermediate purity	High purity	Recombinant
Viral infections	++	++++	+/-	-
Allergic reactions	++++	++	+/-	-
Hemolytic reactions	+	++	-	-
Inhibitors	+	+	+/-	+
Volume	++++	++	-	-
Thrombosis	-	+++	+/-	+/-
Immune suppression	+/-	++	+/-	-

Key:

++++ common ++ very rare - not a concern
 +++ rare + extremely rare

consider one single donor plasmapheresis product to limit donor exposure and risk of infection.

Cryoprecipitate. Use for afibrinogenemia, hypofibrinogenemia, dysfibrinogenemia, factor XIII deficiency in situations where FFP is contraindicated due to volume overload concerns. One donor unit contains ~100 units of factor VIII and vWF and ~1500mg of fibrinogen. Cryoprecipitate has a higher risk of HIV and/or hepatitis C transmission relative to FFP and virally inactivated or recombinant FVIII and vWF concentrates.

Fibrogammin P (FXIII concentrate). May be considered for FXIII deficiency.

Fibrin glue. May be used for small to moderate lacerations and dental extraction but this should be done with caution, as it may lead to the development of thrombin inhibitors.

Special Considerations. *Epistaxis.* Epistaxis is common in children and may or may not be related to a disorder of hemostasis. Management should involve the following:

- Place patient in sitting position to decrease venous pressure. If patient cannot sit, place head higher than heart and turn head to the side.
- If possible, flex the patient's neck forward with the chin touching the chest.
- Firmly compress the lower part of the nose for 20 minutes.
- If bleeding continues, reassess location of compression. If bleeding recurs, reapply pressure for 20 minutes.
- If the patient has a known hemostatic disorder, administer specific treatment as outlined previously.
- Patient should avoid blowing the nose for at least 12 hours to avoid dislodging the clot.
- Consider nasal packing if bleeding is persistent and profuse. If packing is applied, consider humidification or nasal saline sprays to prevent drying of the nasal mucous membranes. Also consider broad-spectrum antibiotics with good coverage for skin flora in immune-compromised patients. Packing may be left in place for up to five days.
- If the area of bleeding can be identified and is circumscribed, consider cauterization.

- If significant blood loss occurs, consider transfusion of PRBC.
- Obtain a basic hemostasis laboratory evaluation and consider evaluation by an ear, nose and throat specialist.

Menorrhagia. The most frequent cause of menorrhagia in children is dysfunctional uterine bleeding, but these can frequently be exacerbated by an undiagnosed underlying mild or moderate vWD. Some studies have shown that up to 20% of women with menorrhagia may have vWD.¹⁶ Immediate treatment should be aimed at controlling bleeding and stabilizing the patient. High-dose estrogen until bleeding ceases followed by a taper then regular oral contraception is usually helpful. If the patient has a diagnosed bleeding disorder, specific treatment should be administered. Patients with significant bleeding may need PRBC transfusion. An evaluation for vWD should be initiated in patients with menorrhagia. Oral contraceptives can increase serum vWF levels and in mild vWD, this is often adequate to allow for allow coagulation.

ITP. Most cases of ITP resolve spontaneously and never need intervention. If the platelet count is less than 10,000/mm³, treatment with IVIG 0.8–1 g/kg should be considered. The dose may be repeated after 24 hours. A response should be expected in 24–48 hours. Other treatment options include steroids, plasmapheresis, rituximab, and other chemotherapeutics. Before administering steroids, careful evaluation should be done to exclude leukemia as a cause of thrombocytopenia. Avoid platelet transfusions, as these are likely to be destroyed by the underlying immune process.

Patients with ITP rarely bleed, but if central nervous system bleeding or other major bleeding occurs, an emergent splenectomy and continuous platelet infusion should be considered.

CNS complications. These can be devastating and range from subtle cognitive defects to debilitating strokes. This underscores why any suspected CNS bleed must be treated promptly.

Hemarthrosis. In addition to acute hemarthrosis, which is characterized by significant swelling, warmth, erythema, pain, and a decrease in range of motion that improves with treatment with specific factor, chronic hemarthrosis occurs in target joints, resulting in persistent limitation in the range of motion with little swelling or erythema. Chronic hemarthrosis may require splinting, prophylactic factor treatment, and physical therapy.

Some long-term complications are treatment-related and range from the development of inhibitors to line infections and HIV infection. **Table 9** shows relative risks of certain complications of various factor replacement products. The risk of transmitting viral infections such as hepatitis, slow viruses, and HIV has continuously decreased as highly purified and recombinant factor replacement products have become the mainstay of treatment.

Summary

A bleeding child can present a diagnostic and management challenge. When confronted with an acutely bleeding child,

immediate management should involve airway management, breathing, and circulation. A detailed assessment to determine the source and extent of bleeding as well as patient and family history should guide further management. Because the possible etiologies of bleeding in children are diverse, resulting in a period of several weeks to arrive at a specific diagnosis, management should be based on probable etiologies. Whenever a life threatening bleed such as CNS bleeding is suspected, treatment should be prompt and not delayed for diagnostic studies. Regional hemophilia treatment centers have specialists available around the clock to assist in the management of hemophilia patients. Having a high index of suspicion for disorders of hemostasis not only helps in establishing specific diagnosis and in appropriate management in case of trauma or surgery, but can avoid unnecessary procedures such as hysterectomies for excessive menorrhagia. Anyone diagnosed with hemophilia should be referred to a comprehensive hemophilia treatment center. With appropriate management, children with bleeding disorders can lead normal lives with few complications.

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CME Questions

21. All patients with hemophilia have a family history of hemophilia.
 - A. True
 - B. False
22. Below what level of FVIII would you expect a prolonged aPTT?
 - A. 75%
 - B. 30%
 - C. 50%
 - D. None of the above
23. Females can develop hemophilia through the following mechanism(s):
 - A. Daughter of a carrier mother and affected father
 - B. Turner's syndrome daughter of a carrier mother
 - C. Extreme lyonization in daughter of a carrier mother and normal father
 - D. All of the above
24. Which of the following statement(s) is/are true about hemophilia?
 - A. Recombinant factor products are the preferred choice for treatment.
 - B. Hemophilia is an expensive disease to treat.
 - C. Always get radiographic images to confirm bleeding before treatment.
 - D. Intramuscular injections are not a concern.
 - E. A and B
25. Which of the following statement(s) is/are true about von Willebrand disease?
 - A. Its severity varies widely.
 - B. It is frequently inherited in autosomal dominant mode.
 - C. It is frequently the underlying or contributing cause of menorrhagia in females.

- D. It can often be treated with DDAVP.
E. All of the above
26. Management of epistaxis should include tilting the head backwards so blood does not flow out of the nostrils.
A. True
B. False
27. Which of the following babies are at risk for hemorrhagic disease of the newborn secondary to vitamin K deficiency?
A. Babies born at home
B. Babies of mothers on anti-convulsant therapy
C. Babies of vegetarian mothers
D. All of the above
E. Only A and B
28. Oral contraceptives are contraindicated in girls with von Willebrand disease.
A. True
B. False
29. Which of the following is/are possible long-term complication(s) of hemophilia?
A. Hemarthrosis

- B. Cognitive deficits
C. HIV infection
D. Hepatitis C
E. All of the above
30. Which of the following statements about ITP is/are true?
A. Patients may present with petechiae.
B. Significant bleeding is rare.
C. Mainstay of treatment is platelet transfusion.
D. In most children, ITP resolves spontaneously and never recurs.
E. A, B, and D

Answers: 21. B, 22. B, 23. D, 24. E, 25. E, 26. B, 27. E, 28. B, 29. E, 30.E

CME Instructions

Physicians participate in this continuing medical education program by reading the article, using the provided references for further research, and studying the questions at the end of the article. Participants should select what they believe to be the correct answers, then refer to the list of correct answers to test their knowledge.

To clarify confusion surrounding any questions answered incorrectly, please consult the source material. After completing this activity, you must complete the evaluation form that will be provided at the end of the semester and return it in the reply envelope provided to receive a credit letter. When your evaluation is received, a credit letter will be mailed to you.

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The CME objectives for *Pediatric Emergency Medicine Reports* are to help physicians:

- Quickly recognize or increase index of suspicion for specific conditions;
- Describe the epidemiology, etiology, pathophysiology, historical and physical examination findings associated with the entity discussed;
- Correctly formulate a differential diagnosis and perform necessary diagnostic tests;
- Apply state-of-the-art therapeutic techniques (including the implications of pharmacologic therapy discussed) to patients with the particular medical problems discussed;
- Provide patients with any necessary discharge instructions.

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PEDIATRIC Emergency Medicine Reports™

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Bleeding Disorders

Causes of Thrombocytopenia

DISORDERS OF DECREASED PRODUCTION	DISORDERS OF INCREASED DESTRUCTION	OTHERS
<ul style="list-style-type: none"> Bone marrow failure syndromes Hereditary Marrow infiltration Chemotherapy-induced Radiation therapy-induced 	<ul style="list-style-type: none"> ITP DIC vWD type IIB Heparin-induced TTP/HUS NAIT Mechanical destruction 	<ul style="list-style-type: none"> Hypersplenism Drug-induced Gestational Infection-related Hemophagocytosis

Abbreviations: ITP—idiopathic thrombocytopenic purpura; DIC—disseminated intravascular coagulation; vWD—von Willebrand Disease; TTP—thrombotic thrombocytopenic purpura; HUS—hemolytic uremic syndrome; NAIT—neonatal alloimmune thrombocytopenia

Common Bleeding Disorders and Basic Laboratory Findings

DISORDER	PLATELET COUNT	PT
Thrombocytopenia	Low	Norr
Platelet dysfunction	Normal	Norr
Hemophilia	Normal	Norr
FVII deficiency	Normal	Prol
vWD	Usually normal	Norr
Vitamin K deficiency	Normal	Prol
Liver disease	Low to normal	Prol
DIC	Low	Prol
Vascular anomaly	Normal	Norr

Key: DIC – disseminated intravascular coagulation; I aPTT – activated partial thromboplastin time

Drugs Associated with Thrombocytopenia

ANTIPLATELET AGENTS	ANTIMICROBIALS	CARDIOVASCULAR AGENTS	NEUROPSYCHIATRIC AGENTS	OTHER
Anagrelide	Amphotericin	Amiodarone	Carbamazepine	Gold
Abciximab	Ampicillin	Captopril	Chlorpromazine	Heparin
Eptifibatid	Isoniacid	Digoxin	Diazepam	
Ticlopidine	Rifampin	Hydrochlorothiazide	Haldoperidol	
Tirofiban	Methicillin	Procainamide	Lithium	
Acetaminophen	Piperacillin	Quinidine	Methyldopa	
Diclofenac	Sulfisoxazole		Phenytoin	
	Bactrim			

Note: This list is not exclusive and includes only common drugs. When confronted with thrombocytopenia of unclear etiology, it is important to always review the patient's medications for possible effects on platelet count.

Adapted from Deloughery T. Hemorrhagic and thrombotic disorders in the intensive care setting. In: Kitchens C, Alving BM, et al, (eds). *Consultative Hemostasis and Thrombosis*. Philadelphia: W.B. Saunders Co.;2002:493-513.

Dosing Guidelines for Hemophilia

TYPE OF BLEED	RECOMBINANT FVIII DOSE FOR HA	RECOMBINANT FIX DOSE FOR HB	OTHER RECOMMENDATIONS
Life-threatening (CNS, airway, hip, GI, retroperitoneal)	50 units/kg; treat suspected CNS bleed prior to imaging	100 units/kg; treat suspected CNS bleed prior to imaging.	<i>CNS:</i> If + CT, continuous factor infusion, 2–4 units/kg/hr for 10–14 days <i>Eye:</i> Consult ophthalmologist <i>Airway:</i> Maintain <i>Compartment syndrome:</i> Consider decompression
Iliopsoas	50 units/kg bolus, then 25 units/kg Q 12 for 10–14 days	80 units/kg bolus, then 40 units/kg QD for 10–14 days	Hospitalize; strict bed rest
Hemarthrosis	25 units/kg bolus; repeat dose in 24 hrs, then Q 48 hrs for 1 week	40 units/kg bolus, then Q 48 hrs for 1 week	RICE, radiographs, splinting
Mucocutaneous (mouth, tooth extraction, epistaxis, menorrhagia)	20 units/kg bolus; Amicar 100–200 mg/kg loading dose, then 50–100 mg/kg Q6 for 10 days	30 units/kg bolus; Amicar 100–200 mg/kg loading dose, then 50–100 mg/kg Q6 for 10 days	Epistaxis: Pressure for 15–20 minutes; consider nasal packing or cautery
Traumatic hematuria	50 unit/kg bolus; maintenance IVF; no Amicar	100 units/kg; maintenance IVF; no Amicar	Bed rest; consider continuous factor
Lacerations requiring sutures	20 units/kg bolus prior to sutures	30 units/kg prior to sutures	Ice and pressure
Pre-operative management depends on procedure and complications	50 units/kg 1 hr prior to surgery; consult hematologist for intra- and postoperative management	100 units/kg 1 prior to surgery; consult hematologist for intra- and postoperative management	Complete blood count, FVIII level, FIX level, and inhibitor titers

Key: RICE—rest, ice, compression, elevate; QD—every day; hr—hour; IVF—IV fluid; CNS—central nervous system; CT—computed tomography; GI—gastrointestinal

Dosing Guidelines for vWD

Life-threatening (CNS, airway, hip, GI, retroperitoneal)	50 units/kg; treat suspected CNS bleed prior to imaging	<i>CNS:</i> If + CT, continuous infusion 3 units/kg/hr for 10-14 days <i>Eye:</i> Consult ophthalmologist <i>Airway:</i> Maintain <i>Compartment syndrome:</i> Consider decompression
Hemarthrosis, muscle, or significant subcutaneous	25 units/kg DDVP for responders	RICE, consider splinting
Lacerations	25 units/kg or DDAVP for responders prior to sutures	Compress to control bleeding; ice
Mucocutaneous (mouth, tooth extraction, epistaxis, menorrhagia)	25 units/kg or DDAVP for responders Amicar 100–200mg/kg loading dose, then 50–100mg/kg q 6hrs for 10 days	Epistaxis: Pressure for 15-20 minutes; consider nasal packing or cautery
Traumatic hematuria	50 units/kg, then consider continuous infusion at 2 units/kg/hr if severe	Bed rest; no Amicar; consider steroids
Pre-operative management depend on procedure and complications	50 units/kg 1 hr prior to surgery, or DDAVP for responders	Consult hematologist for intra- and postoperative management

Key: RICE—rest, ice, compression, elevate; QD—every day; hr—hour; DDAVP—desmopressin; IVF—IV fluid; CNS—central nervous system; CT—computed tomography; GI—gastrointestinal

Characteristics of Hemophilia Bleeding Based on Severity

Aspect	Mild (5%–40%)	Moderate (2%–5%)	Severe (1%)
Age of onset	Usually after 3 yrs	Usually before 2 yrs	Usually first yr
Musculoskeletal bleeding	Joint muscle bleeding rare, except in major trauma	Joint/muscle bleeding with minor trauma; "target joints" rare	Spontaneous joint/muscle bleeding; "target joints" frequent
Postoperative bleeding	Asymptomatic, but wound oozing or hematomas may occur	Wound hematomas common	Brisk bleeding common
Neonatal complications	Usually none	Post circumcision bleeding may occur; ICH is rare	ICH, post-circumcision bleeding may occur
ICH	Very rare, but may occur with significant trauma	May occur	Relatively common; occurs in 3% of pts
Bleeding with dental extraction	Frequently occurs	Commonly occurs	Almost always occurs
Development of inhibitors	Very rare; < 0.5% of pts	Rarely occurs; ~3% of pts	Very common; ~20% of pts
Response to DDAVP	Two- to three-fold level increase	~10 % increase in level	No response

Key: DDAVP – desmopressin; ICH – intracerebral hemorrhage; pts – patients; yr – year
A "target joint" is a joint that has experienced repeated bleeds and is at high risk of re-bleeding.

Adapted from: Dimichelle DM. Hemophilia A. In: Goodnight SH, Hathaway WE (eds), *Disorders of Hemostasis and Thrombosis, A Clinical Guide*. 2nd ed. New York: McGraw Hill; 2001:1127–1139.

Clinical Manifestation of Disorders of Fibrogen Based on Severity

SEVERITY	CLINICAL MANIFESTATIONS
Afibrinogenemia	Severe, lifelong hemorrhagic diathesis.
Hypofibrinogenemia	Mild bleeding e.g. menorrhagia, post-operative bleeding, etc.
Dysfibrinogenemia	Ranges from asymptomatic to mild bleeding to thrombosis

Complications of Various Factor Replacement Products

COMPLICATION	PRODUCT			
	FFP or cryo-precipitate	Intermediate purity	High purity	Recombinant
Viral infections	++	++++	+/-	-
Allergic reactions	++++	++	+/-	-
Hemolytic reactions	+	++	-	-
Inhibitors	+	+	+/-	+
Volume	++++	++	-	-
Thrombosis	-	+++	+/-	+/-
Immune suppression	+/-	++	+/-	-

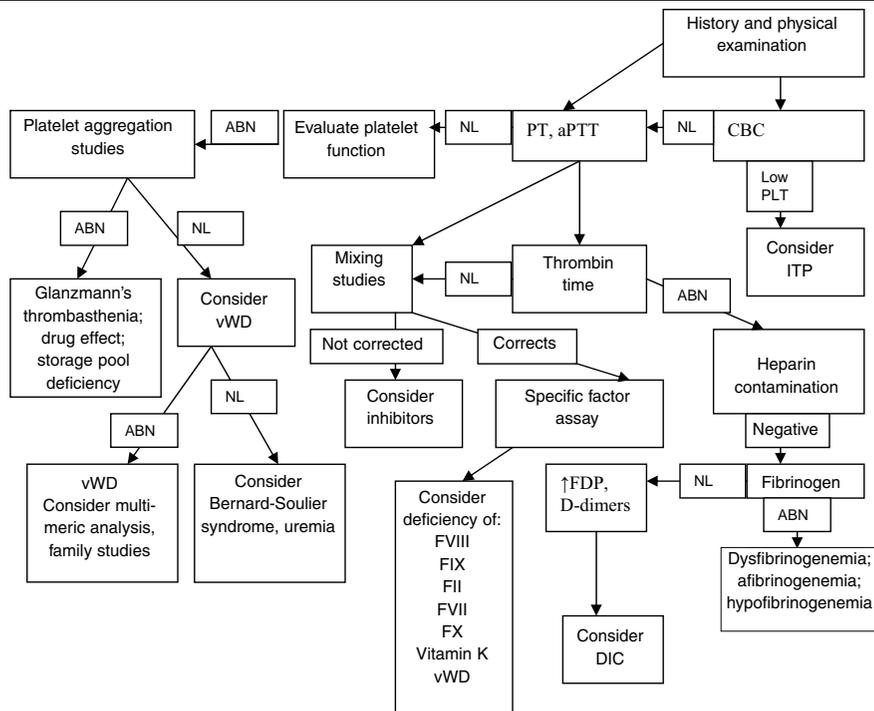
Key:
++++ common ++ very rare - not a concern
+++ rare + extremely rare

A Hemostasis Questionnaire: History to Obtain from Patient or Parent

- extensive (larger than a quarter, indurated), located where trauma is unlikely, and unexplained by minor injury? Are petechiae ever seen? Is there gum bleeding?
- If prolonged bleeding after laceration or surgery:* Has there been prolonged (hours) or recurrent bleeding after lacerations (cuts, oral injury), surgery (circumcision, skin biopsy, tonsillectomy), tooth extractions? Poor wound healing? List all operations and significant trauma.
- If epistaxis or menorrhagia:* Has there been prolonged and heavy menstrual bleeding or severe recurrent epistaxis? If so, was anemia present or need for iron therapy or transfusion?
- If soft tissue or joint hemorrhage:* Is there a history of unusual hematomas or unexplained arthritis or joint swelling?
- Has there been hematemesis, melena, hematuria, or hemoptysis without obvious cause?
- Family history:* Has any blood relative had a problem with excessive bleeding as noted in the questions above?
- General health:* Is there evidence of a disorder known to be associated with a bleeding tendency (e.g., liver disease, renal disease, connective tissue disorder, leukemia, SLE, malabsorption syndrome, myeloproliferative disorder, amyloidosis)? Is there evidence for abuse or self-inflicted injury?
- Drugs or medication:* Has aspirin, an antibiotic, or warfarin been taken in the last 10–14 days? Has vitamin K been used? History of transfusion?

Adapted from Goodnight SH, Hathaway WE (eds). Evaluation of bleeding tendency in the outpatient child and adult. In: *Disorders of Hemostasis and Thrombosis, A Clinical Guide*. 2nd ed. New York: McGraw Hill; 2001: 52-60.

Suggested Decision Tree for the Evaluation of a Child with a Bleeding Disorder



Note: Bold captions denote findings. Arrows point to recommended actions or possible diagnosis.

Key: PT–prothrombin time; aPTT–activated partial thromboplastin time; F–factor; CBC–complete blood count; PLT–platelet count (normal white blood count and hemoglobin are assumed); vWD–von Willebrand disease; FDP–fibrin degradation products; DIC–disseminated intravascular coagulation; NL–normal; ABN–abnormal; ITP–idiopathic thrombocytopenic purpura

Adapted from Hastings CA, Lubin BH. Blood. In: Rudolph AM, Kamei RK(eds), *Rudolph's Fundamentals of Pediatrics*, 2nd ed. Norwalk, CT: Appleton & Lange. 1998: 441-490

Supplement to *Pediatric Emergency Medicine Reports*, March 2009: "Clinical Presentation, Evaluation and Management of Bleeding Disorders in Children." Authors: **Beng R. Fuh, MD**, Assistant Professor of Pediatrics, Department of Pediatrics, Division of Pediatric Hematology and Oncology, Brody School of Medicine at East Carolina University; and **Ronald M. Perkin, MD**, Professor and Chairman, Department of Pediatrics, Brody School of Medicine at East Carolina University. Peer Reviewer: **Afshin Ameri, MD**, Associate Professor of Pediatrics, Director of Pediatric Comprehensive Hemophilia Program, Medical College of Georgia, Augusta.

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Burns may range from a minor injury from a brief contact with hot water to a life-threatening, devastating injury. Burns may be obvious or subtle depending on the mechanism and type of force producing the injury. The early recognition and aggressive management of even the smallest burn makes a significant impact on the outcome of each individual patient, especially in terms of function.

Acute care providers need to be able to aggressively resuscitate a major burn victim and at the same time recognize small burns that may benefit, either based on location or type of burn, from management by a specialist at a burn center. Being aware of burn center resources and appropriate utilization of this available expertise facilitates an optimal outcome in an acutely burned patient.

— The Editor

Introduction

Fires and related burn injuries are a major issue in health care. The U.S. Fire Administration data shows that in 2006, 3,245 civilians lost their lives as the result of fire.¹ There were 16,400 civilian injuries that occurred as the result of fire; 81% of all civilian fire deaths occurred in residences, and 106 fire-fighters were killed while on duty. Direct property loss due to fires was estimated at \$11.3 billion. In 524,000 structural fires, there were 2,705 deaths and 14,350 injuries, resulting in \$9.6 million dollars of direct loss.^{2,3} The U.S. Fire Administration/National Fire Data Center report on fatal fires estimated that there were 3,300 fatal fires that claimed 3,380 civilian lives (86% involved single fatalities, 14% involved multiple fatalities).⁴ Seventy-four percent of fatal fires occurred in structures; 94% of these were on residential properties. The leading cause of fires that resulted in

Optimizing Outcome in the Adult and Pediatric Burn Patient

Author: **Sidney Miller, MD, FACS**, Professor of Surgery; Director, The Ohio State University Burn Center, The Ohio State University College of Medicine University Hospitals, Columbus.

Peer Reviewer: **Carl Menckhoff, MD, FACEP, FAAEM**, Associate Professor, Department of Emergency Medicine, Medical College of Georgia, Augusta.

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UMDNJ Robert Wood Johnson Medical School
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fatalities was arson (27%), followed by smoking (18%). Smoke alarms were either not present or not functional in 63% of residential fires.

The Agency for Healthcare Research and Quality (AHRQ) of the Department of Health and Human Services outcomes data for 2005 for burn injuries in the United States shows 40,687 hospital discharges.⁵ The mean length of stay was 7.1 days, mean charges were \$41,000, and the in-hospital mortality rate was 2.4%. This represents \$1.67 billion in health care cost annually for the management of patients with burns. Of this care, 28.7% was provided under private insurance; however, Medicare and Medicaid paid for 42% of care, and this government expenditure represents \$709 million. Additionally, uninsured patients, whose costs are passed on to other insurers, represented 15% for \$245 million; however, this figure does not represent the entire uninsured group, as many burn patients in most states qualify for Medicaid because of the magnitude of their burn injuries.

Thermal burns may result from contact with flames, hot liquids, hot surfaces, and other sources of intense heat; chemical burns and electrical burns may also occur. In addition, mass casualties and disasters, explosions, and fires can cause a variety of serious injuries, including burns. Prevention and planning are vital; the public must understand how to behave safely in mass casualty and fire situations and to comprehend basic principles of first aid for burn victims, as immediate care can be lifesaving. The Centers for Disease Control and Prevention (CDC) indicates that only 60% of Americans have

an escape plan, and of those, only 25% have practiced it.⁶ The CDC estimates that smoke alarms cut the chances of dying in a fire in half.⁶

This article discusses the initial evaluation and management of the burn patient and summarizes the current approach to these patients.

Initial Evaluation and Management of the Burn Patient

The initial evaluation and management of the burn patient frequently establishes the path for hospitalization and ultimate outcome. Decisions made during the first few hours after injury can have long-term effects on both the functional and cosmetic outcomes.

The goals in the initial management of the burn patient are:

1. A thorough evaluation, as with any trauma patient;
2. Evaluation and management of any traumatic associated injuries that might produce life-threatening hemorrhage, such as fractures, intra-abdominal hemorrhage, and chest trauma;
3. Appropriate evaluation of the status of the burn wound;
4. Determination of the appropriate management course for the patient;
5. Differentiation of whether hospital or ambulatory care is most appropriate; and
6. Initiation of appropriate hospital care, if needed, or arrangement for follow-up care.

The first step in the treatment of any burn is to stop the burning process. All clothing should promptly be removed and a complete examination performed, making sure to include the patient's front and back. Attention should be paid to safeguard the safety of the health care provider as well as the victim, especially in the case of chemical injuries.

The mechanism of the injury is important in the evaluation and management of the burn patient. A patient burned in a closed space has a high possibility for an inhalation injury. If the patient was burned in a motor vehicle crash or explosion, associated traumatic injuries must be considered. Scald burns differ depending on the source. Boiling water can quickly run off the patient, tending to burn less deeply than hot grease, which adheres to the skin and retains the heat as it slowly cools.

An initial primary survey should be performed to determine the presence of any life-threatening injuries. The primary patient survey includes the "ABCs" of airway, breathing, and circulation. The airway must be assessed and established as patent. A history of being burned in a closed space, facial burns, or soot in the oropharyngeal area suggests a possible inhalation injury. Chest x-rays and blood gases are of little use during this early time period; however, if there is delay in establishing the airway until the patient has obvious evidence of respiratory distress, such as wheezing, or severe respiratory distress, oral intubation may not be possible and a cricothyrotomy or tracheostomy may need to be performed.

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Associate Publisher: Coles McKagen
Managing Editor: Allison Weaver
Director of Marketing: Schandale Kornegay

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If signs of an inhalation injury are present and there is a question as to the need for intubation, a bronchoscopy can be performed to assess the status of the airway and, if needed, an endotracheal tube can be inserted. Experience has shown that early placement of an endotracheal tube may avoid a very difficult surgical airway later. The largest tube possible should be placed, at least 7.0 or higher in adults, and in general any patient who requires intubation should be bronchoscoped. Findings on bronchoscopy are prognostic of the extent of the injury and help identify patients who might be extubated early.⁷

Once the airway has been established, attention is turned to the patient's breathing. The lungs must be inflating and the chest fully expanding. Occasionally, a leathery, full-thickness burn encircling the chest will limit excursion of the chest wall, impairing full expansion of the lungs. In such cases, a chest escharotomy may be needed to allow for proper chest wall expansion. In addition, evidence of possible chest trauma, including rib fractures, hemopneumothorax, tension pneumothorax, or flail chest, must be sought and effectively treated.

Any uncontrolled hemorrhage must then be identified and managed. Circulation is supported by the administration of appropriate intravenous (IV) fluids. The amount is based on the burn size estimate and the patient's weight. Two large-bore IVs should be started in the most readily accessible locations; while effort should be made to avoid insertion through any burn area, if patient has extensive burns IVs may need to be started through the burned areas. The patient with a major burn will need to have a Foley catheter inserted so that the adequacy of the fluid resuscitation can be monitored. The "D" in the "ABCDEs" of initial evaluation signifies examining for any disability. The "E" stands for exposing the patient and monitoring the environment, which should be warm to avoid hypothermia in the cold environment of most emergency departments. Initial management should include assessment of any associated injuries, if present. Early deaths in burn patients are usually due to one or more associated injuries. The burn injury itself rarely produces severe hypotension and shock except in extreme circumstances, and other sources of undetected hemorrhage or internal injuries must be sought.

Once the airway is established, the patient is breathing adequately, and access to and support of the circulation is established, the secondary survey is performed. The secondary survey encompasses a complete head-to-toe evaluation of the patient. Only after all of the immediate life-threatening problems have been managed during the primary survey can a complete history and physical examination be performed.

After the primary and secondary trauma surveys have been completed, attention can be turned to the burn wound. The extent of the burn wound must be determined, as well as any associated medical conditions that might adversely affect the patient's outcome. Generally, patients with major burns, which the American Burn Association (ABA) defines as

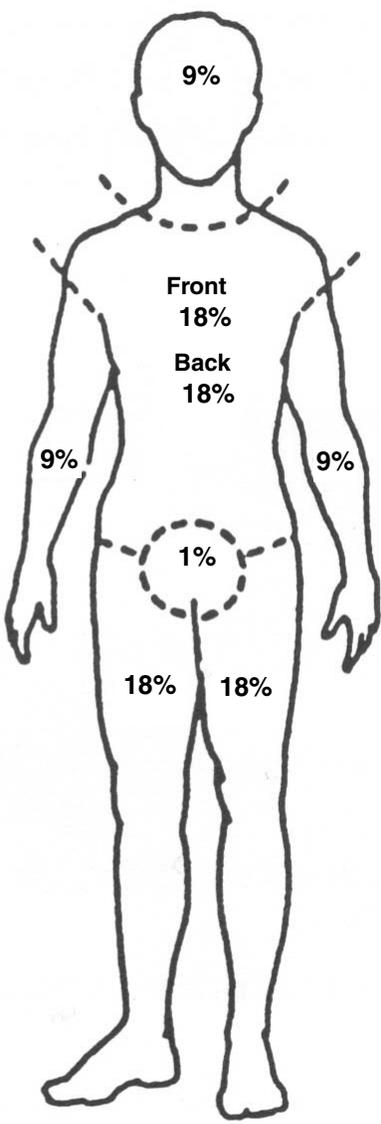
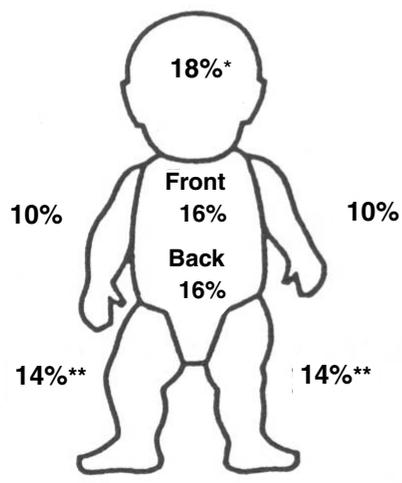
greater than 20% of the body surface area (or greater than 10% in the young and elderly), will require treatment for the effects of burn-wound shock.⁸

The depth of the burn wound is dependent on the temperature and duration of contact of the burning agent. The burn injury rarely is uniform in depth and frequently will have a central area of maximal damage with skin necrosis (zone of necrosis) that is surrounded by a zone of sluggish blood flow referred to as the zone of stasis. More peripherally is the zone of hyperemia, with increased blood flow secondary to the systemic response to the burn injury. In the central zone of necrosis, cellular damage is irreversible and a variety of toxic substances and electrolytes are released into the general circulation. Because of capillary dilation and the local inflammatory response to these released cytokines, fluid and serum proteins are lost from the intravascular space into extravascular space (the "third-space effect"), leading to hypovolemia, hypoalbuminemia, and hypotension. In the zone of stasis, the relatively sluggish blood flow produces cellular hypoxia and acidosis that leads to further hypotension and platelet clumping and, ultimately, to cellular necrosis. This is magnified with prolonged hypotension and shock. The zone of hyperemia is characterized by increased blood flow, with a diversion of blood from the central circulation that further decreases blood flow to vital organs. If circulation remains poor for any reason, or if tissue ischemia and necrosis develop in the area of stasis, additional tissue death will occur and the central zone of necrosis will expand. Therefore, the goals of burn resuscitation are to treat overall hypovolemia, maintain the local capillary circulation to the remaining viable tissues, and reestablish blood flow to vital end organs.

The severity of the burn is a combination of the extent and depth of the burn, the mechanism of the injury, pre-existing medical conditions that might complicate or delay wound healing, and associated injuries. The initial assessment of the extent of the burn is best determined by using a body diagram or chart to estimate burn size. Commonly, the Rule of 9s is used because it is easy to remember.⁹ (See Figure 1.) The body surface of an adult is divided into 11 segments of 9% each, or multiples of 9%, with 1% reserved for the perineum. There are two segments for each leg, two each for the anterior and posterior thorax, one for each arm, and one for the head. When using the Rule of 9s in children, 9% is taken from the legs and added to the head for a child up to age 1 year. Each subsequent year, 1% is returned to the legs until, at approximately age 9, the child's head is in proportion to that of an adult's. For smaller or scattered areas, the palm of the patient's hand, which represents approximately 1% of the patient's body surface, is used to estimate burn size. The more detailed Lund and Browder chart can be used to estimate burn size as, particularly in children, it may be more accurate. The use of these two methods is just about evenly divided among burn units. This suggests that there is not a standard for use of either the Rule of 9s or the Lund and

Figure 1. Rule of 9s

* Subtract 1% from head for each year older than 1 year of age
** Add 1/2% to each leg for each year older than 1 year of age



Used with permission: *Advance Burn Life Support Provider Manual*, 2007 ed. American Burn Association: Chicago, IL; 2007:18. Available at: www.ameriburn.org.

lacking, and much research is being performed in this area.

Equally important in the evaluation of the magnitude of the burn injury is estimating the depth of the burn. Burns are described as partial thickness or full thickness. This classification more truly represents the pathophysiologic status of the injury rather than the older first-, second-, and third-degree classification. (See Figure 2.) A partial-thickness burn involves only a portion of the dermis, and those dermal elements such as sweat glands and hair follicles necessary for re-epithelization of the burn wound remain intact. These skin appendages are necessary for healing of the partial-thickness burn and must have their capillary blood supply maintained by adequate resuscitation. If this blood supply is lost, the surviving appendages will die. In addition to inadequate resuscitation, infection can lead to loss of skin appendages and conversion of a partial-thickness to a full-thickness burn. With the partial-thickness burn, the nerve fibers to the dermis also are preserved. Therefore, the partial thickness burn wound is wet, painful, and blanches with pressure. With the full-thickness burn, all dermal elements have been destroyed, and except for those that involve only a very small area, will require skin grafting. The full-thickness burn is dry, leathery, and insensate.

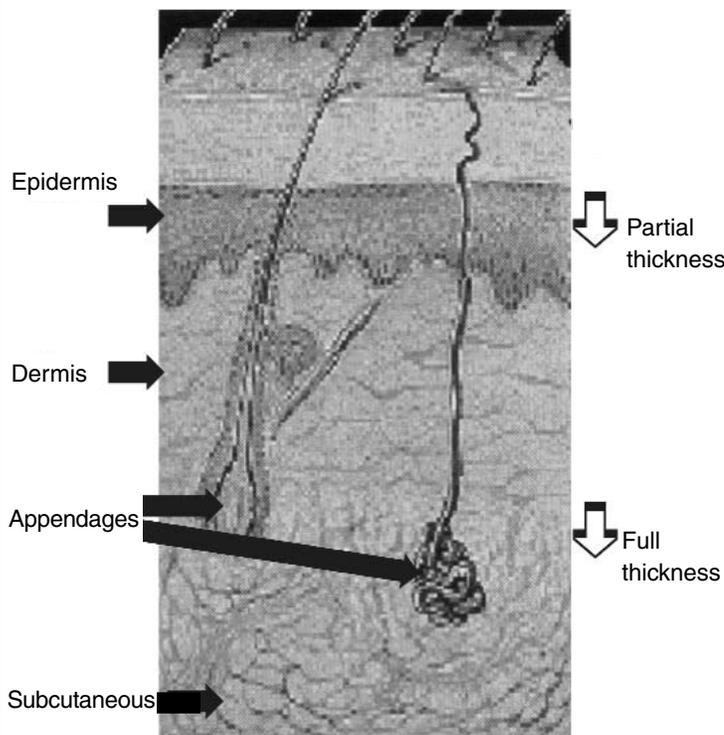
Browder chart, except by facility preference.⁹

It should be noted that these calculations are only estimates, and that superficial burns that are just pink or red, such as sunburn, are not used in the calculation. Only partial-thickness and full-thickness areas of burn are used in estimating the extent of burn.

Since many decisions regarding care, both during and after hospitalization, are based on these "estimates," they should be recorded as accurately as possible and performed by skilled staff. Frequently, hospital staffing and reimbursement are based on these estimates. Unfortunately, objective and accurate methods of estimating burn size and depth are

Following stabilization, a detailed past medical history is essential. Since the major burn injury is still one of the few tetanus-prone injuries, it is important to determine the date of the patient's most recent tetanus immunization. The American College of Surgeons' guidelines should be followed.¹⁰ Tetanus guidelines suggest that if the patient's immunization status is unknown or if the patient has not had the recommended three doses of immunization, both tetanus toxoid and tetanus immunoglobulin should be given. If the patient has had a full three-dose immunization series, tetanus toxoid should be given if it has been longer than five years since the last booster.

Figure 2. Degrees of Burn Thickness



Debate continues regarding the use of nasogastric decompression. Generally, if there will be a delay in transfer of the major burn patient or if the patient will need to be transferred over a long distance, the stomach should be decompressed with a nasogastric tube to avoid the possibility of vomiting and aspiration while en route. Gastric ileus, however, can usually be completely avoided by instituting tube feedings or an oral diet within four to six hours of the burn injury, when possible.

Associated illnesses have a significant impact in the physiologically compromised burn patient. The patient with pre-existing cardiac or renal disease may have additional difficulties with the large fluid requirements of a major burn. The diabetic patient or the patient on corticosteroids is more prone to infections. Additionally, the diabetic patient frequently has peripheral microvascular disease leading to poor wound healing.

Resuscitation of Patients with Major Burns

The primary objectives of fluid resuscitation are to maintain capillary circulation to the potentially viable skin and to support the circulation to vital organs. The estimate of the burn size is used to determine a starting point for resuscitation. Resuscitation is started with crystalloid, usually lactated Ringer's solution, which most closely resembles intravascular serum lost into the extracellular space. The most com-

Table 1. Parkland Formula for Fluid Resuscitation in Burn Patients

1ST 24 HOURS AFTER BURN OCCURS:

- Crystalloid / lactated Ringer's solution
- $4 \text{ mL} \times \text{kg body weight} \times \% \text{ total body surface area burned}$
- 1/2 volume given in first 8 hours; second half given over next 16 hours
- Monitor urine output

monly used formula to determine initial fluid replacement is the Parkland formula, which estimates the IV fluid needs during the first 24 hours after the burn injury as $[4 \text{ mL crystalloid} \times \text{the percentage body surface area burned} \times \text{weight in kg}]$. (See Table 1.) Generally, half of this estimated amount is given over the first eight hours after the injury and half during the next 16 hours, but the rate of fluid administration needs to be adjusted to maintain a urine output of 30–50 mL per hour in adults (1–1.5 mL/kg in children). Fluid overload is associated with increased cardiac strain, and cerebral and pulmonary edema is as harmful as hypovolemia. The balance between adequate fluid resuscitation and minimizing overload is critical — and challenging to achieve.

Ongoing monitoring is very important. Patients with normal mental status should remain alert and oriented and have an appropriate glomerular filtration rate as reflected by an adequate urine output.

Ventilated patients frequently require monitoring of arterial blood gases, typically through an arterial line. Initial resuscitation is started with two large-bore peripheral lines that are usually replaced with a central line at the burn center. Routine use of Swan-Ganz catheter is not recommended, however, as the mechanical trauma across the tricuspid valve can lead to nearly always fatal acute bacterial endocarditis.⁷

The hematocrit, serum osmolality, and serum sodium are routinely monitored. These three tests are additional indicators of the adequacy of fluid resuscitation. Ideally, the hematocrit should be less than 55%, the osmolality less than 350 mOsm, and the serum sodium less than 155 mEq/liter. By maintaining the appropriate electrolyte levels and urine output, adequate fluid resuscitation for the burn patient is assured, and fluid overload can be avoided.

Burns of Special Concern

The overwhelming majority of burns seen are thermal in nature. Electrical and chemical burns, however, are unique and provide very special challenges to the health care provider.

Electrical Injuries. Electrical injuries may be deceptively subtle injuries; there can be tissue damage from the passage of the electric current as well as burn from the ignition of clothing or other materials. Internal injury is a major concern with electrical injury, since the electrical current will pass

Table 2. American Burn Association Criteria for Referral to Burn Center

Table 3. American Burn Association Criteria for Hospitalization of Burn Patient

- Partial-thickness burns of greater than 10% of the total body surface area
 - Burns that involve the face, hands, feet, genitalia, perineum, or major joints
 - Third-degree burns in any age group
 - Electrical burns, including lightning injury
 - Chemical burns
 - Inhalation injury
 - Burn injury in patients with preexisting medical disorders that could complicate management, prolong recovery, or affect mortality
 - Any patients with burns and concomitant trauma (such as fractures) in which the burn injury poses the greatest risk of morbidity or mortality.
 - If the trauma poses the greater immediate risk, the patient's condition may be stabilized initially in a trauma center before transfer to a burn center.
 - Burned children in hospitals without qualified personnel or equipment for the care of children
 - Burn injury in patients who will require special social, emotional, or rehabilitative intervention
- Source:* American Burn Association, Chicago, IL.

- 20% or greater total body burn
- 10% or greater burn of child or older adult
- 5% or greater full-thickness injury
- Burns to the hands, feet, face, or perineum
- Burns of the eyes or ears
- Suspected inhalation injury
- Electrical and chemical burns
- Patients with pre-existing illness
- Patients with associated injuries

through the entire body and can produce difficult-to-detect internal injuries. This passage of electrical current has multiple effects. Electric current can cause contraction and fibrillation of both skeletal and cardiac muscle, destruction of cell membranes, thrombosis of blood vessels, and coagulation necrosis of tissues. The violent contraction of the muscles caused by the passage of the current can produce fractures that may go undetected in the early resuscitation period.

Injured muscles discharge electrolytes and myoglobin into the bloodstream, which have the potential to produce further damage, and released potassium can produce cardiac arrhythmias; cardiac monitoring is mandatory. Myoglobin released from the damaged muscle mechanically plugs the renal tubules and can lead to renal failure. In patients who have myoglobinuria, a higher-than-usual urine output is desired to flush the myoglobin from the renal tubules. These patients have a port-wine-colored urine in the emergency department, and IV fluids must be administered at a rate to produce a urine output of 100–150 mL/hour until the urine grossly clears of myoglobin. Occasionally, an osmotic diuretic may be given to increase urine output and further flush out the myoglobin.

The passage of the electrical current also produces heat, which is produced by the resistance of the various tissues to

the passage of the electrical current. Bone has the highest resistance to the passage of the electrical currents; therefore, a large amount of heat is produced in the bone and the surrounding muscle with no apparent injury to the overlying skin. Necrotic muscle needs to be fully debrided, as it is an excellent medium for bacterial colonization and infection. Amputation is a frequent sequela of extensive electrical injuries.

Chemical Injuries. Chemical burns, like thermal burns, usually involve just the skin. The initial management is dilution by continuous showering for a prolonged period after the injury. Attempts to neutralize the chemical can have an adverse effect and produce further tissue injury. The mixture of a strong acid and a strong base produces an exothermic reaction with further heat production and tissue damage. Knowing the actual chemical producing the injury is important, as many chemicals, particularly industrial chemicals, will require specific treatments or have significant systemic effects when absorbed through the skin. Hydrogen fluoride, a commonly used agent in glass and metal etching, bonds to the subcutaneous tissues and continues to produce damage until neutralized with calcium gluconate. If the injury occurs at work, it is important that any involved chemicals are identified, and that the container of the chemical is brought with the patient to the emergency department for proper identification.

Indications for Hospitalization of the Burn Patient

The American Burn Association has established guidelines for referral of burn patients to tertiary care facilities.⁸ (*See Tables 2 and 3.*) Usually, any patient with a burn involving greater than 20% of the total body surface should be hospitalized. In the young and old, who are less tolerant of burn injuries, a total body burn of 10% warrants hospitalization, and any patient with more than a 5% full-thickness burn should be referred and managed with primary excision, usually within five days of the injury. This is euphemistically referred to as the “5/10/20 rule” of burn care.

Other indications for hospitalization include complicating medical conditions such as diabetes, heart disease, and asso-

ciated injuries. These affect or delay recovery or place the patient at a higher risk. Because of the special needs of patients with significant or circumferential burns involving the perineum, hands, feet, and face, they should be considered for hospitalization even though the burn size might be relatively small. Due to their functional importance, burns of the hands and feet are critical and their management should take place in a burn center, where personnel have experience in caring for these injuries.

Nonsurgical Care of the Burn Patient

The majority of burn patients can be managed with non-operative care. Generally, smaller burns of less than 10% of the body surface area that do not involve critical areas such as the face, hands, feet, or perineum can be managed on an outpatient basis. The three issues that should be addressed in the management of these patients are wound management, pain control, and follow-up care. Wound management is probably the easiest of the three to deal with. Since the early 1970s, effective topical antibiotics for the management of the burn wound have been readily available. Silver sulfadiazine (SSD) cream is most commonly used, as it is painless on application, easy to remove, covers a broad range of skin surface organisms, and is relatively inexpensive. SSD, however, has been shown to delay wound healing and must be changed at least once or twice daily until the wound is healed.¹¹ The wound should be covered with an absorbent dressing that is held in place with roller gauze and/or an ace bandage. Over the past several years, a number of newer silver-containing wound care products have come on the market. Most of these are designed to be applied and left in place for up to a week while the wound heals underneath. Once they have stuck to the wound, the wound usually can be left open to the air. Because the twice-daily dressing changes with SSD are eliminated, pain can usually be controlled with acetaminophen, aspirin, or other NSAID. Elevation of the extremities will aid with both swelling and pain control, and the majority of these outpatients do not need narcotic pain medication.

Smaller burns, generally less than 10% of body surface area, usually can be treated on an outpatient basis. Blisters should be left intact. Those blisters that have opened should be debrided. The wounds should be cleansed with mild soap and water twice daily. Usually, facial burns can be left exposed and covered with a thick layer of triple-antibiotic ointment. The usual topical creams are very drying for the face, head, and neck areas, but can be used on other areas of the body. A well-applied absorbent dressing should be positioned and usually can most easily be held in place with elastic bandages. Limb elevation not only helps to control edema, but also is an important aspect of pain management. Outpatient care can be coordinated with the regional burn center for addressing any questions or concerns. Appropriate follow-up is important to the management of these outpatient burns and frequently is encountered as an area of concern. Many general practitioners neither have the expertise nor the

time necessary for dressing changes in the office for other than the smallest-size burns. Many hospitals, however, have wound care centers that can comfortably manage these outpatients, even those with larger-surface-area burns. The regional burn center will almost always have an outpatient component and frequently is the best site for the management for these patients.

Pain Management

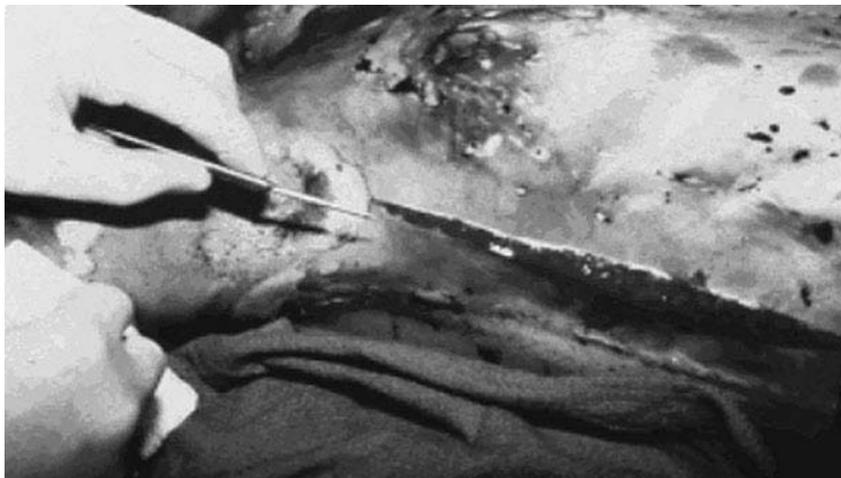
Pain management is a major issue with the burned patient. During the early resuscitative phase of patient management, blood flow to the skin is disrupted for a number of reasons that alter the absorption of intramuscular or subcutaneously delivered pain medication. The neuron-hormonal response to the loss of intravascular fluid volume during the shock phase redirects blood flow to the central core to support blood flow to vital organs. Additionally, subsequent edema development as fluids leak from the intravascular space to the extravascular space during this shock phase (third-space effect) hinders the absorption of intramuscularly administered medication. Judicious use of IV pain medication is indicated during this period.¹² Pain medications may be titrated to control pain and avoid adverse reactions. Pain scores and response to medication should be recorded and monitored.

After the initial burn shock period, the magnitude of the individual patient's pain response may differ. In the very young and very old, pain management can be challenging, as the response of patients in these two groups can be quite variable. With the major burn patient who is admitted to the hospital, pain management takes two forms. The first is management of background pain; this type of pain is generally constant pain that is present until wound closure has occurred, which might be several weeks to months. Once the patient is able to tolerate oral pain management, a long-acting narcotic such as methadone is started and titrated to manage this background pain. Shorter-acting narcotic and NSAIDs also are used to supplement the effects of the long-acting agents.

Episodic pain occurs with specific activities such as dressing changes and wound or extremity manipulation during the necessary physical and occupational therapy required during recovery. Control of this episodic pain is quite variable and dependent on many factors, including how stoic the patient is, past experiences with pain management or drug use, or pre-existing, chronic, painful conditions that might compound the management episodic pain during this particular period of time. A variety of pain management methodologies are employed, including small doses of IV medication, oral pain medications, and the liberal use of anxiolytic agents. The goal is to use the smallest amount of pharmacologic agents as possible to control this episodic pain. Alternative pain management, including music therapy, imagery, and virtual reality, has also been of value.¹⁰⁻¹²

The hallmark of pain management in the outpatient setting is a well-applied wound dressing and limb elevation to

Figure 3. Chest Escharotomy



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decrease dependent edema during the early phase after the burn injury. NSAID agents or acetaminophen are the mainstay of outpatient management. Again, the needs of any particular patient will be influenced by his or her past experience with pain and drug usage, although oral narcotics may be needed for breakthrough pain.

Surgical Care of the Burn Patient

Escharotomy. During the early post-burn period, the patient's extremities may appear pale, feel cool, and exhibit poor capillary refill due to shunting of blood from the periphery to the central core. Once proper resuscitation is initiated and the peripheral circulation is re-established, there is release of a variety of inflammatory mediators. These mediators lead to transudation of intravascular fluids into the extravascular space with the loss of fluids and proteins, producing local swelling. As this swelling occurs in the area of the full-thickness burn, with its rigid and unyielding overlying eschar, compartment syndrome can develop and the adequacy of the peripheral circulation must constantly be monitored. In patients with circumferential burns of the extremities, this is especially concerning and the practitioner should be vigilant for signs of developing compartment syndrome. While usually a late sign, any loss of pulses would certainly suggest vascular compromise.

An escharotomy performed with a scalpel or by electrocautery through the full-thickness burn to the subcutaneous fat allows this rigid tissue to expand and releases the constricting pressure on the underlying vessels. While this should be a painless procedure, as it is usually through an area of full-thickness burn, there is frequently brisk venous bleeding that can be managed with a bulky dressing and elevation of the extremity. **Figure 3** shows a chest escharotomy performed to improve ventilation in patients with circumfer-

ential chest burns. **Figure 4** is a chart of escharotomy sites for extremities and the chest. With extremity escharotomies, peripheral pulses should be re-established, and with chest escharotomies, airway pressures should decrease. Local or regional anesthesia usually is not necessary.

Wound Coverage. The classical management of burns for centuries involved waiting for the eschar to separate naturally from the underlying subcutaneous tissue, with subsequent skin grafting, one of the oldest surgical procedures. This eschar separation was due to subeschar infection and liquefaction of the nonviable tissue by bacterial proteolytic enzymes. Sepsis was common, as the bacteria trapped beneath the eschar frequently gained access to the systemic circulation. Since the 1960s, effective topical antibiotics that actively penetrate the eschar have been available. Their use, however, delays eschar

separation. Delayed eschar separation results in lengthened patient hospitalization and recovery.

In 1970, Janzekovic published on a series of patients treated with early excision and grafting.¹³ The rationale was that the earlier the dead skin was removed, the faster the wound would heal. Excision done within the first 3–4 days resulted in an excellent take of skin grafts, lower sepsis rates, and shorter hospitalizations. If the patient enters the burn unit late or with burn wounds already heavily colonized, skin grafting may result in loss of the skin graft and should be postponed until the bacteria count is acceptable.

Primary excision has become the standard in the management of burn patients, with removal of all nonviable burned skin either by using a dermatome, scalpel, or electrocautery. Excision can be tangential, sequential, or full-thickness. Once the eschar is removed, the next important stage of surgical management of a burn patient is wound coverage.

There are several alternatives to achieve wound closure, the most efficient being to excise the burn wound and suture the skin closed. This is an acceptable method for some small burns and frequently is overlooked. Small burns managed in this fashion usually heal with minimal scarring, and the procedure can be performed on an outpatient basis.

With burns of less than 20% of the body surface area, there is usually an adequate supply of donor sites for skin grafting and wound coverage. Small full-thickness burns might be allowed to heal on their own, as the base frequently is too small to support a skin graft. Larger full-thickness burns and deep partial-thickness burns that are estimated to take longer than three weeks to heal require skin grafting. Burns requiring skin grafting usually are managed by early total excision, although wound coverage sometimes can become an issue because of the lack of available donor sites. With smaller-area burns requiring skin grafting, cosmetic and

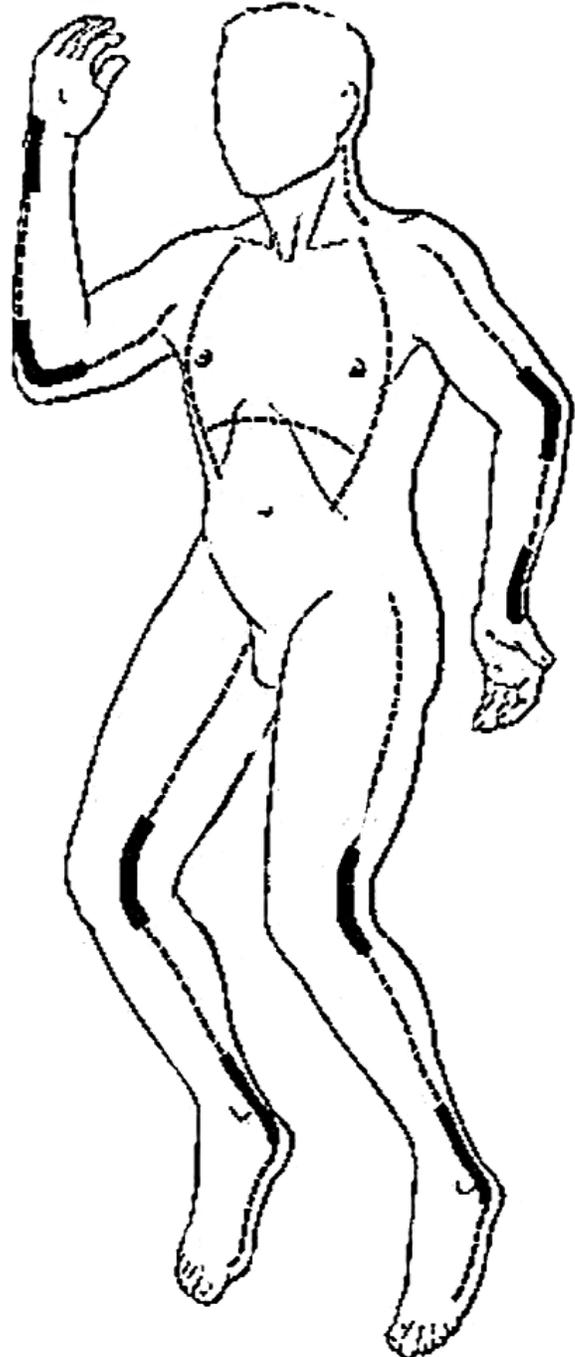
functional areas such as hands and faces are addressed first. With larger-area burns, patient survival is the first priority, and skin grafting follows certain other priorities. The wound should be closed as quickly as possible to decrease the risk of colonization of the necrotic skin and the chance of systemic sepsis. Wound closure is accomplished most easily in large, flat surfaces such as the chest, abdomen, and anterior upper and lower extremities. With early excision and good skin graft adherence, the total burn size is decreased gradually to a smaller, nonlethal size. Coverage of the burn wound with autografts depends on the extent and location of the wound and the amount of donor skin available. Grafts may be applied as “sheet” grafts, whereby a strip of skin is removed from a donor site and transferred without alteration. Sheet grafts are more durable and produce better cosmesis than meshed grafts, which is particularly beneficial for coverage of areas exposed to shearing forces or the environment such as hands, neck, arms, and face.

With large wounds, however, the available donor sites frequently are insufficient to cover the excised area. An early attempt to expand the amount of available skin was made by using “pinch” grafts, in which many small, fingertip-size grafts were used to cover the burn wound. In 1964, Tanner described a meshing device that cuts small holes in the graft and allows for expansion of the harvested skin graft to allow for coverage of large wounds.¹⁴ The expansion ranges from 2:1 up to 9:1, and experimental expansions of up to 100:1 have been studied in the laboratory.^{15,16} The 9:1 expansion is very difficult to work with and infrequently used; 2:1 to 4:1 are used most commonly. The pattern of the meshed skin graft will remain when the graft heals; therefore, every effort is made to avoid the use of meshed grafts on the face, neck, and hands. Donor sites generally heal in 10–14 days and can be re-harvested three or four times. Because donor sites resemble partial-thickness burns, they are usually painful, but with the newer donor-site dressings this pain usually subsides in 3–4 days.

As the application of primary excision has expanded, additional methods of expanded mesh grafting have been developed. One of these is a cultured skin substitute called cultured epithelial autograft (CEA).¹⁷ CEA is used when large, total body surface area burns (generally greater than 50%) require coverage and patients do not have enough donor skin available. A small piece of unburned skin is excised and keratinocytes are grown in a laboratory. These grafts take several weeks to grow, during which time patients are at risk for infection. Although cultured keratinocytes provide more material to cover the patient’s wounds, they are less durable than autografts, as they contain no dermal elements. Several additional models are being developed in the laboratory that either grow epithelial grafts earlier on a fibroblast matrix to increase durability, or in which liquid keratinocyte cultures are applied in a spray fashion in the early post-burn period. The keratinocytes spray is under clinical evaluation.

Aggressive, early excision has stimulated a market for a

Figure 4. Escharotomy Sites



The dashed lines indicate the preferred sites for escharotomy incisions. The solid segments of the lines emphasize the importance of extending the incisions across involved joints.

Source: Emergency War Surgery, Second United States Revision of The Emergency War Surgery NATO Handbook. *Operational Medicine*. United States Department of Defense;2001:Fig. 16. Available at www.brooksidepress.org/Products/OperationalMedicine/DATA/operationalmed/Manuals/NATOEWS/figures/fig16.html. Used with permission.

variety of biological and biosynthetic products. These products include allogeneic cadaver skin, human allogeneic dermis, and the “artificial” skin Integra™ (Integra Lifesciences Corp., Plainsboro, NJ), which is a biosynthetic two-part construct of shark cartilage and bovine collagen with an outer silicone sheet.¹⁸ With proper care, once Integra™ “takes,” at about 3–4 weeks the outer silicone sheet is removed and a skin graft is applied. It is unclear whether the Integra™ survives long term or merely serves as a strut for the in-growth of the patients’ own fibroblasts. Motion and infection are the primary causes of Integra loss, and the vacuum-assisted closure device has proved beneficial in improving Integra™ assimilation.

The biologic dressings also include porcine or other species xenografts. Amnion is an excellent biologic dressing, but its use has been limited in recent years due to the potential risks of hepatitis and HIV transmission.¹⁹

A variety of biosynthetic dressings have been developed to temporarily cover the excised full-thickness burn or the partial-thickness wound and promote wound healing. Temporary wound coverings provide a protective barrier either while donor sites heal for future harvesting, until Integra™ is ready for grafting, or for primary coverage of partial-thickness burns. One major advantage of these temporary dressings is that many can be left in place over the partial-thickness wound until it heals, eliminating painful, twice-daily dressing changes. Many of the new synthetic wound coverings have characteristics of biologic coverings, and most are some form of synthetic matrix such as silicone with biologic components such as bovine collagen. Many also have some form of silver ions applied to them, which have antibacterial effects.

The ultimate goal of surgical management is to achieve a functional, durable, and cosmetically acceptable skin surface. Early surgery in the burn patient involves removal of the full-thickness burn and its replacement either with the patient’s own skin (autografts) or one of a number of temporary biological or biosynthetic dressings. Patients with large burns will require multiple operations to cover their wounds and may require multiple reconstructive procedures to achieve maximal cosmetic and functional outcomes.

Recently, some patients undergoing elective cosmetic surgery after effective weight-reduction surgery have offered to donate “extra” skin to burn centers, either from good Samaritan desires or for imagined financial gains. These donations are very problematic, and most burn and tissue centers have refused such offers.

Summary

The management of the acute burn injury provides the framework for patient survival and sets the entire course of their hospitalization. Assessments made during this period will have a far-reaching effect on the resultant disability or function that the patient might experience.

The burn patient always must be approached with a com-

plete assessment, just like any other trauma patient. This includes the ABCs of airway, breathing, and circulation, and the recognition of associated life-threatening injuries. A past medical history of any illnesses that might complicate the healing of the burn wound should be obtained. The burn wound needs to be properly assessed for depth and extent to determine the course of treatment. IV fluid replacement needs are calculated from the size estimate.

Hospitalization and the need for referral to a regional burn center should be determined using American Burn Association guidelines. (See Tables 2 and 3.) The surgical management of these patients has become quite complicated, with early excision being effectively applied to improve patient survival, shorten hospital stays, and improve patient outcomes.

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CME Questions

1. The Centers for Disease Control and Prevention estimates that all of the following are true *except*:
 - a. Only 60% of Americans have a fire escape plan.
 - b. Only 25% of those who have a fire escape plan have practiced it.
 - c. Smoke alarms cut occupants' chances of dying in a fire in half.
 - d. Malfunctioning smoke alarms are the major cause of home fire deaths.
2. Burn patient management includes all of the following *except*:
 - a. Thorough head to toe evaluation
 - b. Evaluation and management of any associated injuries
 - c. Full-body CT scan to exclude other injuries
 - d. Determination of the appropriate management course
 - e. Initiation of appropriate hospital care, if needed, or arrangement for follow-up care
3. The three zones of burn injury are:
 - a. Zones of stasis, necrosis, and hypoperfusion
 - b. Zones of stasis, hyperemia, and necrosis
 - c. Zones of necrosis, hyperperfusion, and stasis
 - d. Zones of necrosis, stasis, and hypoperfusion
 - e. Zones of necrosis, stasis, and hypoperfusion
4. Early bronchoscopy and intubation are suggested for which of the following?
 - a. All major burn patients
 - b. Patients burned in a closed space with facial burns or soot in the oropharynx
 - c. Patients with abnormal chest x-rays
 - d. Patients with abnormal blood gases
 - e. Only for patients in acute respiratory distress
5. The "ABCDEs" of burn management include all of the following *except*:
 - a. A is for airway
 - b. B is for breathing
 - c. C is for circulation
 - d. D is for disability
 - e. E is for escharotomy

6. "Rule of 9s" refers to:
 - a. Determining burn sizes of nine body areas
 - b. Using 9 mL IV Ringers for each percent body burn
 - c. Dividing the body into 11 areas of 9% body surface area each
 - d. Dividing the burn size by 9 to determine fluid needs
7. Partial-thickness burns (as opposed to full-thickness burns) are all of the following *except*:
 - a. Red
 - b. Wet
 - c. Painful
 - d. Leathery
8. The Parkland formula for fluid resuscitation is:
 - a. Crystalloid usually given as LR 7 mL/kg/% body surface area burn
 - b. Crystalloid usually given as LR 4 mL/kg/% body surface area burn
 - c. Crystalloid usually given as LR 2 mL/kg/% body surface area burn
 - d. Crystalloid usually given as D5NS 4 mL/kg/% body surface area burn
9. The usual cause of early renal failure in electrical injury is:
 - a. Shock
 - b. Sepsis
 - c. Myoglobinuria
 - d. Multi-system organ failure (MSOF)

CNE/CME Instructions

Physicians and nurses participate in this continuing medical education/continuing education program by reading the article, using the provided references for further research, and studying the questions at the end of the article. Participants should select what they believe to be the correct answers, then refer to the list of correct answers to test their knowledge. To clarify confusion surrounding any questions answered incorrectly, please consult the source material. **After completing this activity, you must complete the evaluation form provided and return it in the reply envelope provided in order to receive a letter of credit.** When your evaluation is received, a letter of credit will be mailed to you.

CNE/CME Objectives

- Upon completing this program, the participants will be able to:
- a.) discuss conditions that should increase suspicion for traumatic injuries;
 - b.) describe the various modalities used to identify different traumatic conditions;
 - c.) cite methods of quickly stabilizing and managing patients; and
 - d.) identify possible complications that may occur with traumatic injuries.

10. American Burn Association indications for hospitalization of burn patients include:
- Great than 20% total body burn
 - Greater than 5% full-thickness burn
 - Any suspected inhalation injury
 - All electrical and chemical burns
 - All of the above

“The presentation of the information is excellent. ED Legal Letter emphasizes pitfalls and gives the conclusions . . .”



Answers: 1. a; 2. c; 3. b; 4. b; 5. e; 6. c; 7. d; 8. b; 9. c; 10. e

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