



Contemporary Approaches to the Management of Hemophilia A: An Individualized Approach to Care

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- Webinar registration link
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Contemporary Approaches to the Management of Hemophilia A: An Individualized Approach to Care

Part One

Contemporary Approaches to the Management of Hemophilia A: An Individualized Approach to Care

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Provided by ASHP

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Contemporary Approaches to the Management of Hemophilia A: An Individualized Approach to Care

Learning Objectives

- Discuss the epidemiology and pathophysiology of hemophilia A.
- Review the available long-acting factor replacement products, including patient-specific factors to consider with their use.
- Explain established care models and standards and criteria for the care of persons with hemophilia A.
- Describe important hematologic and pharmacologic monitoring parameters to employ when treating patients with hemophilia A.

On average how many unique patients with hemophilia A (not unique patient encounters) do you personally provide care to each month?



- a. Less than 5 patients/month
- b. 6-10 patients/month
- c. 11-15 patients/month
- d. More than 15 patients/month
- e. None – I am not directly involved in patient care

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Hemophilia

- The word Hemophilia is derived from two Greek words
 - Hemo: blood
 - Philia: affection, like
- Inherited or acquired bleeding disorder
 - Deficiency of clotting factor
- Hemophilia Types
 - Inherited Hemophilia A: Factor VIII (FVIII) deficiency (classic hemophilia)
 - Inherited Hemophilia B: Factor IX (FIX) deficiency (Christmas disease)
 - Acquired: most common factor VIII

Centers for Disease Control and Prevention. "What is Hemophilia?" www.cdc.gov/ncbddd/hemophilia/facts.html
Srivastava A et al. *Haemophilia*. 2013; 19:e1-e47.

Epidemiology

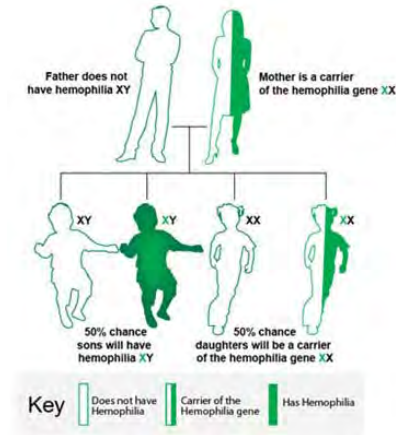
- Average incidence of hemophilia A and B
 - Estimated 1 in 5,032 live male births
 - Estimated 400 babies born every year in the U.S.
- 79% hemophilia A
- Age-adjusted prevalence of inherited hemophilia is 13.4 cases/100,000
 - 10.5/100,000 for hemophilia A
 - 2.9/100,000 for hemophilia B

Soucie JM et al. *Am J Hematol*. 1998; 59:288-94.

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Etiology: Congenital Hemophilia

- X-linked recessive disorder
- Male patients
- Female carriers
- One third have no family history of bleeding

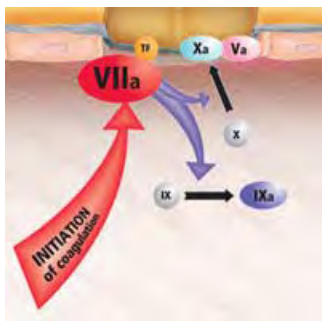


Centers for Disease Control and Prevention. "What is Hemophilia?" www.cdc.gov/ncbddd/hemophilia/facts.html

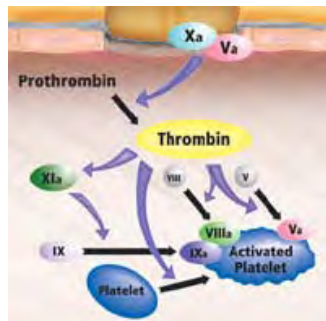
Pathophysiology

Cell-Based Coagulation Model

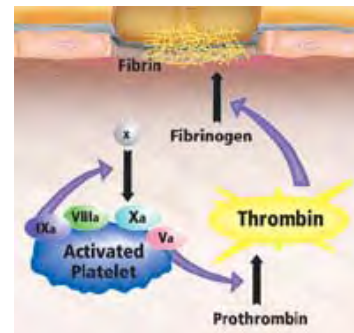
Initiation



Amplification



Propagation

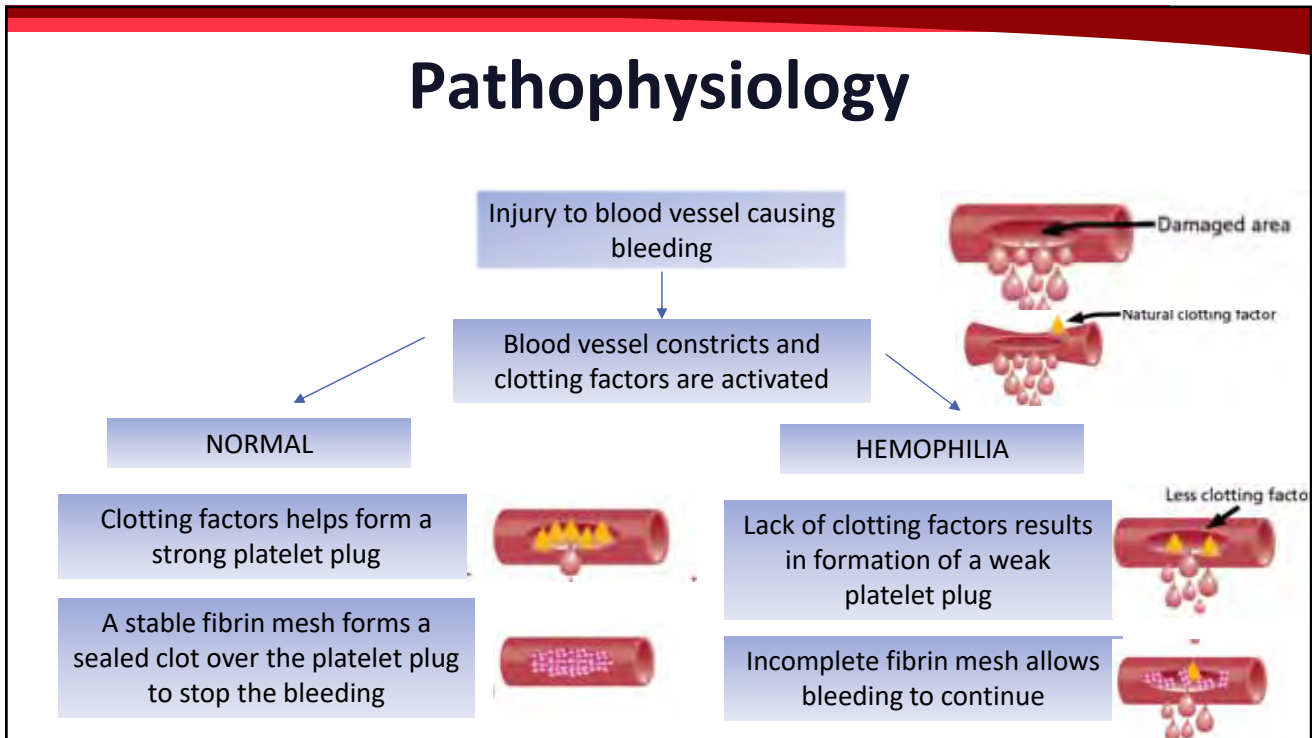


www.anaesthesiauk.com/article.aspx?articleid=100097.

Hoffman M, Monroe MM. *Thromb Haemostat*. 2001; 85:958-65.

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Pathophysiology



Clinical Classification of Congenital Hemophilia

Mild

- Baseline factor level 5-40%
- Rare spontaneous bleeding
- Severe bleeding with major surgery or trauma
- Median age at diagnosis: 36 months

Moderate

- Baseline factor level 1-5%
- Occasional spontaneous bleeding
- Prolonged bleeding with minor trauma or surgery
- Median age at diagnosis: 18 months

Severe

- Baseline factor level <1%
- Spontaneous bleeding
 - Joint, muscles
- Median age at diagnosis: 1 month

Srivastava A et al. *Haemophilia*. 2013; 19:e1-e47.

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Bleeding in Hemophilia A

Bleeding Severity	Bleeding Sites	Approximate Frequency
Serious	Joints (hemarthrosis)	70-80%
	Muscles (deep compartments: iliopsoas, calf, forearm)	10-20%
	Mucous membrane (mouth, gum, nose, genitourinary tract)	
Life-threatening	Intracranial	<5%
	Neck/throat	5-10%
	Gastrointestinal tract	

Srivastava A et al. *Haemophilia*. 2013; 19:e1-e47.

Treatment of Bleeding Episodes

- Initiate treatment as soon as possible
 - Within 2 hours of onset of symptoms
 - Before imaging
- Replacement of the deficient clotting factor
 - Increase FVIII with desmopressin
 - Factor VIII concentrates
 - Intermittent bolus or continuous infusion
- Utilization of bypassing agent (BPA) if inhibitors (i.e., antibodies to clotting factors) present
 - Recombinant activated factor VII (rFVIIa) or activated prothrombin complex concentrate (aPCC)

Srivastava A et al. *Haemophilia*. 2013; 19:e1-e47.

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Factor Dosing in Hemophilia

- Dosed in **units**
 - 1 unit is the amount of factor present in 1 mL of normal plasma
- Current and desired clotting factor level
 - Refer to the World Federation Hemophilia Guidelines for desired level
- Half-life of the factor
 - FVIII - 12 hours
- Recovery of factor level
 - 1 unit/kg of FVIII raises plasma level by 2%

$$\text{Dose} = (\text{desired level} - \text{baseline level}) \times 0.5 \times \text{weight}$$

Srivastava A et al. *Haemophilia*. 2013; 19:e1-e47.

Patient Case Example

- Patient has severe hemophilia A
 - baseline level <1%, Weight 80 kg
- CNS bleed: desired level 100%
- Dose = (100%-0%) X 0.5 X 80 kg
= 4000 units
- Continuous infusion
(50 units/kg X 2) / 24 hr
= 4 units/kg/hr = 320 units/hr
Dose = (desired level – baseline level) X
0.5 X weight

Type of hemorrhage	Desired level (%)	Duration (days)
Joint	40-60	1-2
Iliopsoas and deep muscle		
Initial	80-100	1-2
Maintenance	30-60	3-5
CNS/head		
Initial	80-100	1-7
Maintenance	50	8-21
Gastrointestinal		
Initial	80-100	1-7
Maintenance	50	8-14

Srivastava A et al. *Haemophilia*. 2013; 19:e1-e47.

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Polling Question 1



TA, a 60-kg male, is in the ED with right knee bleeding. He has moderate hemophilia A with a baseline level of 1%.

What would you recommend for his first dose of factor VIII?

- a. 30 units/kg = 1800 units
- b. 40 units/kg = 2400 units
- c. 50 units/kg = 3000 units
- d. 100 units/kg = 6000 units

Dose = (desired level – baseline level) X 0.5 X weight

Type of hemorrhage	Desired level (%)	Duration (days)
Joint	40-60	1-2
Iliopsoas and deep muscle		
Initial	80-100	1-2
Maintenance	30-60	3-5
CNS/head		
Initial	80-100	1-7
Maintenance	50	8-21
Gastrointestinal		
Initial	80-100	1-7
Maintenance	50	8-14

Adverse Effects of Factor Replacement

- Inhibitor development
- Transfusion-transmitted infections
 - HIV
 - Hepatitis C virus

Srivastava A et al. *Haemophilia*. 2013; 19:e1-e47.

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Inhibitors in Hemophilia A

	Severe hemophilia	Mild, moderate hemophilia
Incidence	20-30%	5-10%
Median age of diagnosis	3 years old	30 years old
Pathophysiology	Inactivation of exogenously administered clotting factor	Inactivation of exogenously administered clotting factor, and endogenously synthesized factor VIII
Timing of inhibitor development	Within 150 exposure days to CFC	After intensive CFC exposure during surgery
Bleeding sites	Joint bleed	Mucocutaneous, urogenital, gastrointestinal bleed

CFC=clotting factor concentrate

Srivastava A et al. *Haemophilia*. 2013; 19:e1-e47.

Inhibitors in Hemophilia A: Treatment Strategies

Treatment of bleeding, if present

- Bypassing agents (BPA)
- Recombinant activated factor VII (rFVIIa) or activated prothrombin complex concentrate (aPCC)

Treatment to eradicate the inhibitor

- Immune tolerance induction (ITI)
 - Severe hemophilia
 - High-dose factor VIII 100 units/kg/day
- Immunosuppressants
 - Mild, moderate hemophilia
 - Rituximab, cyclophosphamide, corticosteroids

Srivastava A et al. *Haemophilia*. 2013; 19:e1-e47.

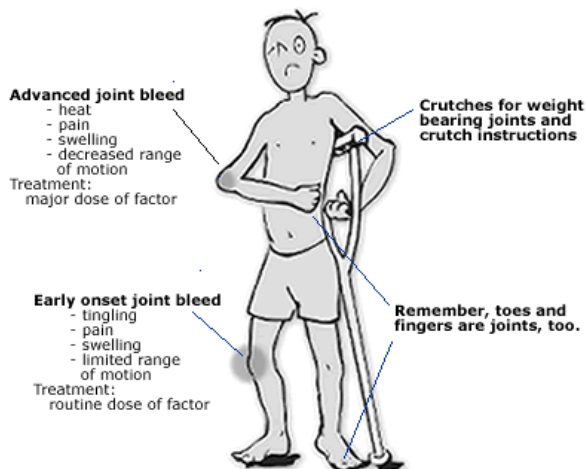
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Treating Bleeding in Patients with Inhibitors

- Activated prothrombin complex concentrate
 - 50-100 units/kg every 8-12 hours, maximum 200 units/kg/day
 - Dose >200 units/kg/day = increased risk of disseminated intravascular coagulation (DIC)
 - Risk of thrombosis
 - Breakthrough bleeding
 - No in vitro assay available to measure hemostatic efficacy
- rFVIIa
 - Severe bleeding
 - 90 mcg/kg every 2 hours until hemostasis achieved, then every 3-6 hours
 - Minor, moderate bleeding
 - 90 mcg/kg every 2 hours until hemostasis achieved

FEIBA [prescribing information]. Lexington, MA: Baxalta US, Inc.; Jan 2019.
Novoseven [prescribing information]. Princeton, NJ: Novo Nordisk, Inc.; Jan 2019.

Hemarthrosis



Rebleeding

- Worsening of symptoms on treatment or within 72 hours after stopping treatment

Target joints

- 3 or more spontaneous bleeding episodes within a 6-month period

Srivastava A et al. *Haemophilia*. 2013; 19:e1-e47.

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Treatment of Hemarthrosis

- On-demand treatment of acute joint bleeding
- Prophylactic treatment
 - Prevent joint bleeding and destruction
 - Preserve normal musculoskeletal function
 - Factor replacement
 - Short-acting factor concentrates
 - Long-acting (extended half-life [EHL]) factor concentrates
 - Non-factor replacement
 - Emicizumab-kxwh

Srivastava A et al. *Haemophilia*. 2013; 19:e1-e47.

Prophylaxis

Primary

- Before second joint bleeding episode
- Before age 3 years
- Absence of joint disease

Secondary

- After 2 or more joint bleeding episodes
- Before age 3 years
- Absence of joint disease

Tertiary

- After 2 or more joint bleeding episodes
- After age 3 years
- Presence of joint disease

Srivastava A et al. *Haemophilia*. 2013; 19:e1-e47.

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Prophylactic EHL Factor VIII Concentrates

	FDA approval	Half-life (hours)	Prophylactic dosing
AHF, pegylated (Adynovate)	Nov 2015	14.69	40-50 units/kg IV two times weekly
AHF, Fc fusion protein (Eloctate)	June 2014	16.4	50 units/kg IV every 4 days
AHF, pegylated-aucl (Jivi)	Aug 2018	21.4	30-40 units/kg IV two times weekly
AHF, glycopegylated-exei (Esperoct)	Feb 2019	21.7	50 units/kg IV every 4 days

AHF: Antihemophilic Factor (also known as Factor VIII)

Adynovate [prescribing information]. Lexington, MA: Baxalta US, Inc.; November 2018.

Eloctate [prescribing information]. Cambridge, MA: Biogen; January 2017.

Jivi [prescribing information]. Whippany, NJ: Bayer Healthcare LLC; August 2018.

Esperoct [prescribing information]. Plainsboro, NJ: Novo Nordisk, Inc.; 2019.

Prophylactic EHL Factor VIII Concentrates

	Subjects	Dosage (IV)	Efficacy	Safety
AHF, pegylated (Adynovate)	137 subjects Age 12-65 yr	<ul style="list-style-type: none"> •Prophylactic 40-50 IU/kg twice weekly •On-demand 10-60 IU/kg 	<ul style="list-style-type: none"> •90% reduction in ABR •95.9% of bleeding episodes successfully treated with 1 to 2 infusions 	<ul style="list-style-type: none"> •Diarrhea, nausea, headache, and flushing •No inhibitor development
AHF, Fc fusion protein (Eloctate)	165 subjects Age 12-65 yr	<ul style="list-style-type: none"> •Arm 1: Prophylactic 25-65 IU/kg every 3-5 days (n=118) •Arm 2: Prophylactic 65 IU/kg weekly (n=24) •On-demand: 10-50 IU/kg (n=23) 	<ul style="list-style-type: none"> •92% reduction in ABR in arm 1 •76% reduction in ABR in arm 2 •87.3% of bleeding episodes successfully treated with 1 infusion 	<ul style="list-style-type: none"> •Nasopharyngitis, arthralgia, headache, upper respiratory infection •No inhibitor development

ABR: annualized bleeding rate

Konkle BA et al. *Blood*. 2015; 126:1078-85. Mahlangu J et al. *Blood*. 2014; 123:317-25.

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Prophylactic EHL Factor VIII Concentrates

	Subjects	Dose (IV)	Efficacy	Safety
AHF, pegylated-aucI (Jivi)	134 subjects Age 12-65 yr	<ul style="list-style-type: none"> •10-week run-in period: 25 IU/kg two times weekly •>1 bleed: 30-40 IU/kg two times weekly •≤1 bleed: 45-60 IU/kg every 5 days or 60 IU/kg every 7 days 	<ul style="list-style-type: none"> •Pre-study ABR 15.3 •ABR post <ul style="list-style-type: none"> •2x weekly: 1.9 •Every 5 days: 1.9 •Every 7 days: 3.9 •90.6% of bleeding episodes successfully treated with ≤2 infusions 	<ul style="list-style-type: none"> •Nasopharyngitis, arthralgia, back pain, headache, cough, epistaxis •No inhibitor development
AHF, glycopegylated-exei (Esperoct)	186 subjects Age ≥12 yr	<ul style="list-style-type: none"> •Prophylactic 50 IU/kg every 4 days •On-demand 20-75 IU/kg 	<ul style="list-style-type: none"> •Prophylaxis: median ABR 1.33, mean 3.7 •95.5% of bleeding episodes successfully treated with ≤2 infusions 	<ul style="list-style-type: none"> •Nasopharyngitis, arthralgia, musculoskeletal pain, viral infection •1 patient developed inhibitor

Reding MT et al. *J Thromb Haemost.* 2017; 15:411-9. Giangrande P et al. *Thromb Haemost.* 2017; 117:252-61.

Personalized Treatment Considerations

- Patient characteristics
 - Bleeding pattern
 - Joint status
 - Daily activities (level and timing)
 - Adherence to treatment
 - Blood type
- Interpatient variability
 - Pharmacokinetic (PK) profile of the patient's replacement factor

Hazendock H CAM et al. *Blood Reviews.* 2018; 32:265-71.

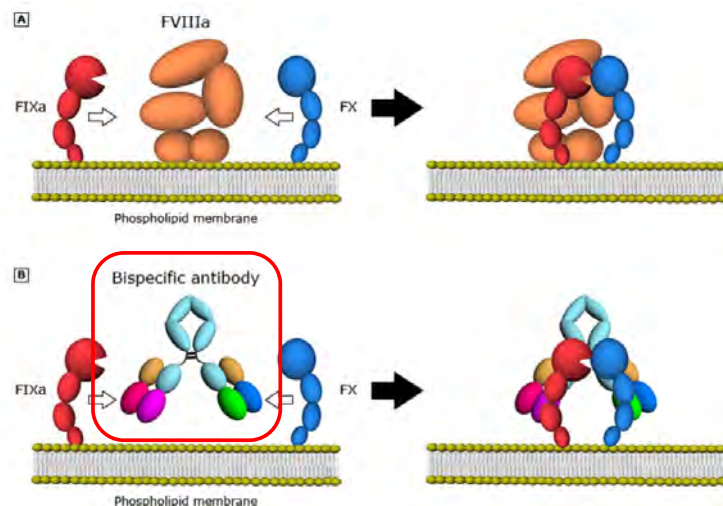
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Candidates for PK-guided dosing

<p>High Bleeders, Low trough levels <u>“Frail or high activity” patients</u></p> <ul style="list-style-type: none"> • Increase dosing to higher trough in order to adjust to patient’s bleeding risk • Optimize regimen according to scheduled activity • Improve adherence 	<p>High Bleeders, High trough levels <u>“Difficult to treat” patients</u></p> <ul style="list-style-type: none"> • Evaluate time of bleeding occurrence with predicted factor levels • Optimize regimen according to scheduled activity • Improve adherence
<p>Low Bleeders, Low trough levels <u>“Well controlled” patients</u></p> <ul style="list-style-type: none"> • Maintain utilization with tailored dosing in order to maintain outcomes 	<p>Low Bleeders, High trough levels <u>“Potential to lower trough levels” patients</u></p> <p>Adjust regimen to target appropriate trough</p> <ul style="list-style-type: none"> • Reduce usage by frequency and/or dosage • Optimize regimen according to scheduled activity

Gringeri et al. *Expert Rev Pharmacoecon Outcomes Res.* 2016; 16:337-45.

Emicizumab-kxwh



Sampei Z et al. *PLOS ONE.* 2013;

<http://journals.plos.org/plosone/article?id=10.1371/journal.pone.0057479>

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Emicizumab-kxwh

- Not affected by existing factor VIII inhibitors and does not cause development of new inhibitors
- Pharmacokinetic properties
 - Linear kinetics
 - Elimination half-life 4-5 weeks
- Pharmacodynamic properties
 - Trough levels ≥ 50 mcg/mL demonstrated a bleeding rate of zero in at least 50% of study participants
- FDA approved in November 2017
 - Routine prophylaxis to prevent or reduce the frequency of bleeding episodes
 - Adult and pediatric patients ages newborn and older
 - Hemophilia A (congenital factor VIII deficiency) with or without factor VIII inhibitors

Yoneyama et al. *Clin Pharmacol Ther.* 2016; 99:Suppl: PI-003.

Emicizumab-kxwh Studies

	Study population	Number of subjects	Efficacy	Safety
Haven 1	Hemophilia A patients with inhibitors	109	Bleeding rate with emicizumab-kxwh was 87% lower than with no emicizumab-kxwh	<ul style="list-style-type: none"> • Most common: injection site reactions • Thrombotic microangiopathy (aPCC use) • Thrombosis
Haven 3	Hemophilia A patients without inhibitors	152	Bleeding rate with emicizumab-kxwh was 96-97% lower than with no emicizumab-kxwh	<ul style="list-style-type: none"> • Most common: injection site reactions

Oldenburg J et al. *N Engl J Med.* 2017; 377:809-18. Mahlangu J et al. *N Engl J Med.* 2018; 379:811-22.

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Emicizumab-kxwh Dosing Pearls

- Loading dose of 3 mg/kg subcutaneous once weekly for 4 weeks
- Maintenance dose (subcutaneous)
 - 1.5 mg/kg once weekly
 - 3 mg/kg every 2 weeks
 - 6 mg/kg every 4 weeks
- Choice of maintenance dose is dependent on healthcare provider, taking patient adherence into consideration
- Discontinue prophylactic use of bypassing agents before starting emicizumab-kxwh
- Prophylactic factor VIII may be continued during the first week of emicizumab-kxwh

Hemlibra (emicizumab-kxwh) [prescribing information]. South San Francisco, CA: Genentech, Inc.; October 2018.

Polling Question 2



Which of the following is true regarding prophylactic factor replacement?

- a. Prophylaxis could be achieved with factor concentrates only.
- b. Prophylaxis prevents joint bleeding and joint destruction.
- c. Prophylaxis is not recommended if joint damage is present.
- d. Prophylaxis with long-acting factor VIII products is no more effective than with short-acting factor VIII products, and should not be recommended.

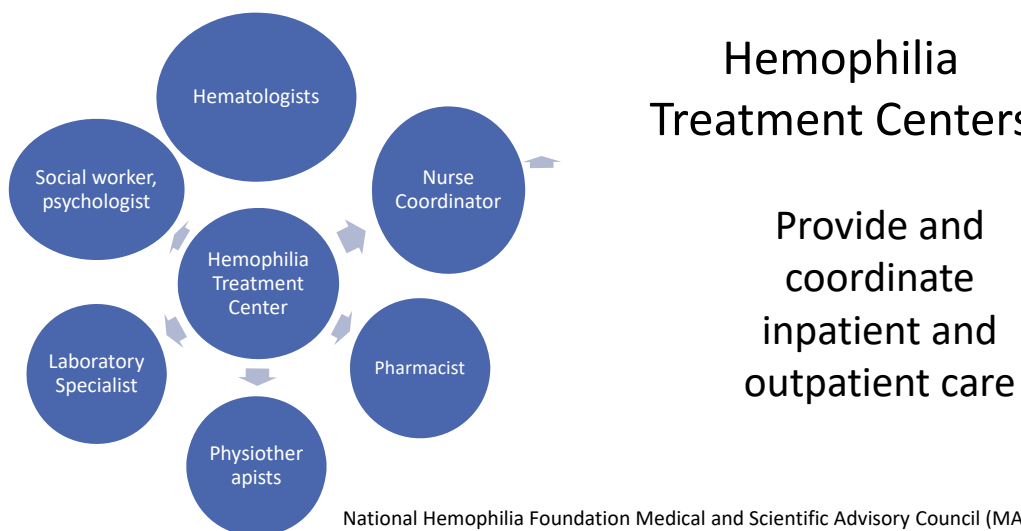
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Established Care Models: Hemophilia Treatment Centers (HTC)

- CDC study on hemophilia patients in 6 states from 1993-2005
 - 2950 subjects followed for average 2.6 years
 - Median age 22 years, 79% hemophilia A, 42% severe disease, 67% HTC
 - Age-adjusted mortality rate of 40.4 deaths /1000 patient-years
 - 65% deaths HIV related
 - Independent risk factors for death
 - HIV/AIDS, liver disease, Medicare/Medicaid
 - Mortality rates 40% lower among patients with HTC care

Soucie JM et al. *Blood*. 2000; 96:437-42.

Standards and Criteria for Care of Patients with Hemophilia



National Hemophilia Foundation Medical and Scientific Advisory Council (MASAC) #132:
www.hemophilia.org/sites/default/files/document/files/masac132.pdf

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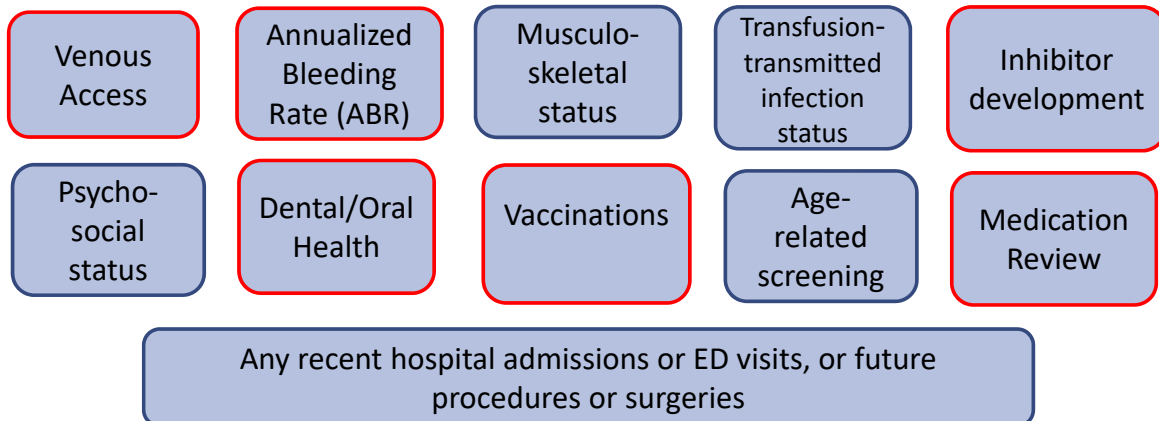
Comprehensive Care Services

- Comprehensive evaluation
 - Medical history, physical examination
 - Nursing evaluation
 - Physical therapy evaluation
 - Psychosocial evaluation
 - Dental evaluation
 - Laboratory testing
- Genetic counseling
- Treatment products
 - Medication access
- Home and self infusion
- Psychosocial services
- Hemophilia carriers
 - Symptomatic
 - Pregnancy

MASAC #132: www.hemophilia.org/sites/default/files/document/files/masac132.pdf

Hemophilia Treatment Centers

Regular evaluations at least every 12 months (adults), 6 months (children)



Srivastava A et al. *Haemophilia*. 2013; 19:e1-e47.

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Hematologic Monitoring Parameters

- Factor activity levels
- Factor inhibitor levels
- Complete blood count
- Transfusion transmitted infection status
 - HIV, hepatitis C virus
- Annualized bleeding rate

Pharmacologic Monitoring

- Medication review
- Assessment of adherence
- Institutional policy on patients with bleeding risk
- Vaccinations

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Medication Review

- Drug-drug interactions
 - Drugs that may increase bleeding risk
 - Antiplatelet agents, anticoagulants
 - Drugs that increase fall risk
- Avoid route of administration that may increase bleeding risk
 - Intramuscular injections
 - Rectal enema or suppositories
- Avoid constipating medications

Pharmacist Role in Adherence

- Education for the patient and caregiver on
 - Medication
 - Medical condition
- Continuous monitoring of patient's prophylactic therapy
 - Breakthrough bleeding episodes
- Monitoring of patient's ability to take own medications correctly and adherence to prescribed therapies
 - Assess administration technique
 - Encourage drug administration log
 - Memory aids
 - Reminder letter/telephone calls for default on refills

World Health Organization. Adherence to long-term therapies; evidence for action. 2003.
www.who.int/chp/knowledge/publications/adherence_full_report.pdf

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Nursing Policy on Patients with Risk of Bleeding

- Observe patient for signs and symptoms of bleeding
- Schedule peripheral blood draw once daily
- Apply firm pressure for 5-10 minutes to all skin puncture sites
- Avoid use of tourniquets and automatic blood pressure cuffs
- Avoid intramuscular injections
- Fall prevention
- Mouth care with oral swabs, soft bristle toothbrush
- Lubricate lips
- Avoid rectal temperatures
- Confirm factor replacement plan in place before procedures/surgery

University of North Carolina Medical Center. Risk of Bleeding Nursing Policy (accessed May 27, 2019).

Vaccination in Hemophilia A

- Protocol for administration of vaccines
 - Fine gauge needle (25-27 gauge) should be used
 - Application of firm pressure for at least 5 minutes to injection site without rubbing
 - Avoid aspirin and NSAIDs
 - Avoid intramuscular injection whenever possible
 - Intramuscular injection given soon after a dose of factor replacement therapy
 - Vaccine administration within 1 day of administration of clotting factor concentrate if patient is on prophylaxis
- Subcutaneous administration of vaccines
 - Pneumococcal polysaccharide (PPSV)
 - Inactivated polio vaccine (IPV)
 - Hepatitis A and hepatitis B virus

MASAC Document #221, www.hemophilia.org/Researchers-Healthcare-Providers/Medical-and-Scientific-Advisory-Council-MASAC/MASAC-Recommendations. Srivastava A et al. *Haemophilia*. 2013; 19:e1-e47.

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Key Takeaways

- When in doubt, treat with factor concentrates first
- Always consider patient-specific factors in factor dosing
- Communication, communication, communication

Practice changes

- Be proactive in counseling patients on bleeding risk.
- Investigate which care models my institution uses for the care of persons with congenital bleeding disorders.
- Read my institution's protocols for the monitoring of patients with hemophilia A.
- Review which factor replacement products are available at my institution.
- Discuss with colleagues how to assess the need for a change in therapy (e.g., development of inhibitors, acute bleeding).
- Educate colleagues on the appropriate dosing of long-acting factors.

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Selected Resources

- Srivastava A et al. *Haemophilia*. 2013; 19:e1-e47.
- National Hemophilia Foundation Medical and Scientific Advisory Committee (MASAC) recommendations.

www.hemophilia.org/Researchers-Healthcare-Providers/Medical-and-Scientific-Advisory-Council-MASAC/MASAC-Recommendations