Etiology

- Hereditary disorder that is X-linked recessive.
- Acquired hemophilia is the development of FVIII inhibitors (autoantibodies) in persons without a Hx of FVIII deficiency.

Usual Treatment

- Desmopressin (DDAVP injection or Stimate nasal spray) whenever possible for mild hemophilia A.
- Treatment includes clotting factor replacement therapy and recombinant FVIII and FIX products; plasma and recombinant factor products are now considered safe and equally effective; and there is no reported seroconversion to HIV, HVB, or HVC.
- Plasma concentrations of deficient factors are maintained at a minimum of 40–70% throughout the periop period; for major procedures, 100% is recommended before surgery and is maintained for 24–48 h.
- Cryoprecipitate is no longer recommended as a treatment alternative.
- Thrombin is produced via alternative pathways; prothrombin complex concentrate and recombinant factor VIIa (NovoSeven) is used in pts with inhibitors to FVIII of FIX.
- Gene insertion therapy shows a promising future.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Pharyngeal bleeding	Often seen in children	Tongue and mouth lacerations	Exam
GI	GI bleeding not common	When it occurs, bleeding can be excessive	Stool exam and endoscopy	Hemoccult, angio
HEME	Anemia, hematoma formation, and bruising	Lethargy, SOB, and skin discoloration	Hematomas	PT/PTT, plt count, bleeding FVIII and FIX assay, gene analysis
GU	Hematuria	Blood in urine		UA, cysto, IVP
CNS	Intracranial hemorrhage	Head trauma, headache, and change in mental status	Any sign or symptom of head injury or trauma	Head CT
MS	Joint hemorrhage Joint deformities Muscle hemorrhage Compartment syndrome Chronic pain	Painful distention of the joint Bruising Restricted movement Narcotic dependence	Hemarthroses Limited ROM Tenderness	Physical exam X-ray

Key References: Franchini M, Mannucci PM: Past, present and future of hemophilia: a narrative review, *Orphanet J Rare Dis* 7:24, 2012; Cabani LM, Ramsey G: Hemostasis and transfusion medicine. In Barash PG, Cullen BF, Stoelting RK, et al, editors: *Clinical anesthesia*, ed 7, Philadelphia, PA, 2013, Lippincott Williams & Wilkins, pp 433–434.

Perioperative Implications

Preoperative Preparation

- Preparation for the care of a pt with hemophilia should include consultation with a hematologist and when available, a hemophilia treatment center.
- Ideally, the anesthesiologist should have experience treating pts with bleeding disorders.
- A comprehensive detailed plan should be in place that outlines the type of hemophilia, factor levels, and dosing strategy for replacement of coagulation factor deficiencies.
- In elective surgery, levels of deficient coagulation factor should be restored to 40–70% of normal before surgery.
- Inhibitor screening and inhibitor assay assessments are essential to preop preparation.
- Adequate quantities of clotting factor concentrates should be available for surgery and the postop course.
- One unit of factor concentrate per kilogram of body weight normally increases the factor concentration by 2%. For factor VIII the half-life is 6 to 10 h, and for factor IX the half-life is 8–16 h, and thus approximately 1.5 U/h per kg of factor VIII or 1.5 U/2 h per kg of factor IX should be given.

Airway

- Care with laryngoscopy to avoid any trauma and thus bleeding to the airway is essential.
- Avoid nasal cannulations (i.e., endotracheal and nasogastric tubes).

Maintenance

- Noninvasive monitoring is optimal; risk/benefit ratio of invasive monitoring vs. site bleeding risk deserves significant consideration.
- Incorporate techniques to minimizing blood loss relative to the procedure.
- Antifibrinolytic drugs such as tranexamic acid are effective as adjunctive treatments for mucosal bleeds and dental extractions.

Adjuvants

- Risk of uncontrolled bleeding detracts from the selection of regional anesthetic technique. However, brachial plexus blocks performed at the axilla without complications have been reported.

Anticipated Problems/Concerns

- Blood bank support for plasma components, if needed, should be included in plans.
- Minimal risk of transmitting hepatitis and AIDS accompanies transfusion of blood components.

Hemosiderosis, Pulmonary

Fredrick Ntumy | Roy G. Soto

Risk

- Classically affects infants and children (80% manifest before 10 y old), but can affect any age group.
- No predilection for males or females.

Worry About

- Restrictive lung disease
- Pulm Htn
- Cor pulmonale
- Alveolar hemorrhage
- Increased need for transfusion secondary to acute and chronic anemia
- Adrenal insufficiency secondary to chronic steroid use

Overview

- Rare disorder of unknown etiology characterized by repeated episodes of intraalveolar hemorrhage and deposition of hemosiderin in alveolar macrophages. The cycle of recurrent hemorrhage frequently leads to the development of pulm fibrosis, pulm Htn, and cor pulmonale. Disease course is variable and can be marked by multiple spontaneous remissions, and the extent of pulm hemorrhage can be massive, leading to early death, or can be clinically insignificant.
- Presents with classically with the triad of hemoptysis, anemia, and pulm infiltrates on CXR.

Etiology

- Unknown; thought to be immune-mediated due to its responsiveness to immunosuppressive therapy
- Associated with immune-mediated disorders such as Goodpasture syndrome, SLE, Heiner syndrome, and Wegener granulomatosis, which can cause diffuse alveolar hemorrhage via immune-mediated mechanisms

Usual Treatment

- Immunosuppression with steroids (IV and PO) and supportive care. Other immunosuppressive agents such as azathioprine, chloroquine, and cyclophosphamide may be tried in steroid-unresponsive pts.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
RESP	Pulm hemorrhage, restrictive lung disease, pulm Htn	Fatigue, weakness, cough, dyspnea, hemoptysis	Tachypnea, pallor, tachycardia, crackles, wheezing, clubbing, growth failure	CXR, PFTs, TTE
CV	Cor pulmonale, ischemia (secondary to anemia and CAD)	Fatigue, tachypnea, exertional dyspnea, cough, angina	Cardiac exam with emphasis on right heart failure	ECG, TTE
ENDO	Adrenal suppression secondary to chronic steroid use; pts may need stress dose steroids			
HEME	Acute and chronic iron deficiency anemia	Fatigue, exertional dyspnea, angina (if CAD)	Pallor of mucous membranes, tachycardia	CBC, iron studies

Key References: Bakalli I, Kota L, Sala D, et al: Idiopathic pulmonary hemosiderosis—a diagnostic challenge, *Ital J Pediatr* 40:35, 2014; Soto RG, Soares MM: Idiopathic pulmonary hemosiderosis in pregnancy: anesthetic implications, *J Clin Anesth* 17(6):482–484, 2005.

Perioperative Implications

Preoperative Preparation

- Evaluate for ongoing alveolar hemorrhage (look for classic signs and symptoms); delay elective surgery in pts with acute disease.
- Assess extent of restrictive lung disease; pt may need PFTs, ABG, and pulm optimization depending on procedure and severity of pt's disease.
- A decrease in vital capacity below 15 mL/kg or the presence of hypercapnia suggest that the pt is a high-risk candidate for pulm compromise.
- Assess degree of anemia and correct as needed to maximize oxygen carrying capacity.
- Evaluate pt for coagulopathy.
- Pts may require stress dose steroids if on chronic immunosuppressive therapy.
- Treat infections.
- Consider postponing elective procedures in setting of alveolar hemorrhage.

Monitoring

- Blood loss (pt may need transfusion)
- Emphasis on ventilation and oxygenation
- Airway pressures

Airway

- Use largest possible ETT for pt to allow for bronchoscopy and pulm toilet in the event of acutealveolar hemorrhage.

Induction

- Be wary of hypotension and the potential for cardiac ischemia in pt with decreased oxygen-carrying capacity and CAD.

Maintenance

- Check Hb/monitor blood loss.
- Avoid high airway pressures (use smaller TVs and/or increase inspiratory time) to avoid barotrauma or pneumothorax.
- Use PEEP.

Extubation

- Use standard extubation criteria.

Adjuvants

- Transfuse blood as needed.

Postoperative Period

- Maintain adequate oxygenation and ventilation.

Anticipated Problems/Concerns

- Acute alveolar hemorrhage

Henoch-Schönlein Purpura

Madhuri S. Kurdi

Risk

- Most common childhood systemic vasculitis; rare in adults.
- Reported annual incidence varies between 10-30 cases per 100,000 in children younger than 17 y and 3.4–14.3 cases per million in adults.
- Mean age of presentation is 6 y; mainly affects children between 4-11 y of age in up to 90% of cases.
- Occurs most commonly in spring; associated with recent URTIs in 90% of cases.
- Cases are reported all over the world; highest incidence is found in Caucasians and lowest in African Americans in North America.

Perioperative Risks

- Morbidity/periop complications increase with abnormal renal function and neurologic/pulm/cardiovascular involvement/emergency surgery.

Worry About

- Problems of concurrent supportive medications (NSAIDs, immunosuppressants, steroids, ACE inhibitors) that the pt may be taking
- Hypoproteinemia due to proteinuria if renal involvement
- Anemia due to hematuria if renal involvement and GI bleeding
- Fluid and lyte imbalance due to N/V and renal involvement

Overview

- HSP is an acute, self-limiting, autoimmune, small vessel childhood vasculitis commonly affecting those of the dermis, bowel wall, and rarely the ureter, myocardium, adrenals, brain, and lungs. Glomerular mesangial hypercellularity with endocapillary proliferation occurs commonly.
- It begins commonly with a nonthrombocytopenic purpuric rash. Arthritis or arthralgia is present in three-quarters of children and approximately 61% adults. GI symptoms occur in up to 85% of children and 48% of adults. Renal involvement is seen in 20–55% of children and approximately 32% of adults. GN is seen in a third of cases and may manifest as isolated hematuria, hypertension, or nephritic/nephrotic syndrome. 1–5% of children and 50% of adults with renal involvement progress to ESRD. Renal failure is the most common cause of death. The disease usually runs its entire course in 4 wk, and many children have no permanent sequelae. Renal symptoms can develop up to 3 mo after initial presentation. The course is complicated in adults.
- HSP is a clinical Dx, and none of the laboratory features are pathognomonic. Palpable purpura plus at least one feature like diffuse abdominal pain/IgA deposition in any biopsy/arthritis/renal involvement suggests the Dx.

Etiology

- Unknown; often triggered by URTI due to respiratory pathogens like group A Streptococcus, methicillin resistant *Staphylococcus aureus*, *Helicobacter pylori*, hepatitis HIV, parvovirus B19, multiple vaccines including H1N1 vaccine, insect bites, drugs like penicillin, quinine, chlorothiazide, food allergies, and malignancy-associated tumor antigens.
- Involves IgA-mediated autoimmune hypersensitivity; the large immune complexes formed face the problem of impaired clearance, settle in the small vessel walls of the affected organs, and trigger an inflammatory response.

Usual Treatment

- Mainly supportive and symptomatic; includes maintenance of adequate hydration, symptomatic pain relief with opioids/NSAIDs, and monitoring for the development of complications
- Short course of low dose oral steroids for those with severe abdominal pain
- High-dose IV corticosteroids, azathioprine, cyclophosphamide, cyclosporine, plasmapheresis, IV immunoglobulins for massive GI hemorrhage/severe proteinuria
- ACE inhibitors for severe nephritis, dapsone for vasculitis, colchicine for skin lesions sometimes
- Renal transplant in ESRD; emergency surgery for acute abdomen due to intussusception/bowel ischemia or perforation