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

Assessment Points

System	Effect	Assessment by Hx	PE	Test
DERM	Cutaneous vasculitis: Petechiae, purpura (common)	Rash	Symmetric palpable nonblanching nontender purpura over extensor surfaces of lower limbs, predominantly buttocks, forearms; trunk and face sometimes Lesions appear in crops Erythematous macular rash sometimes Subcutaneous edema over scalp, hands, feet Blisters and ulcers in adults sometimes	CBC with plt count PBS PT aPTT
GI	Edema and focal hemorrhage of bowel wall and mesentery: Acute abdomen, GI bleed, intussusceptions with an intestinal wall hematoma as the lead point	Colicky, poorly localized abdominal pain (most common) Nausea, vomiting are common, bloody diarrhea, melena, obstipation (less common)	On abdominal palpation—rigidity/distension/guarding/mass	Stool guaiac test for occult blood Abdominal US CT, MRI of abdomen GI endoscopy
GU	Microscopic hematuria, variable grade proteinuria, acute GN, nephrotic syndrome, distal ureteric stenosis, rapidly progressive GN ESRD	Renal colic, pink urine (hematuria), foamy urine (proteinuria), fatigue, swelling around face and eyes, weight gain	Facial/scrotal/penile edema Check BP for Htn	Urine microscopy for RBCs, RBC casts Urine dipstick test for blood and protein BUN Serum Cr, protein; lytes, especially potassium Abdominal US for kidneys, ureter Percentage of cellular crescents on renal biopsy for prognosis indication
MS	Symmetric arthritis/arthralgia of ankle, knee, hip, elbow Cervical joint arthritis rarely	Joint pains/swelling, neck pain	Periarticular swelling, tenderness, erythema, Decreased range of joint movements Restricted neck movements	X-ray/MRI of involved joints X-ray/MRI of cervical spine
CNS	Cerebral vasculitis, myelopathy, subarachnoid hemorrhage, PRES (rarely)	Headache, drowsiness, altered mental status, seizures, stroke	LOC Detailed neurologic examination for paresis, focal deficits, neuropathies	Contrast CT/MRI/MRI angiography brain, spine
CV	Vasculitis: Myocarditis, AV block (rarely), Htn if renal dysfunction	Palpitations Dizziness, syncope, chest pain	Look for bradycardia, ventricular arrhythmias, signs of heart failure	ECG, 2D ECHO, CXR, CT chest
RESP	Interstitial pulm hemorrhage/pneumonia/fibrosis (rarely)	Breathlessness, cough	Signs of respiratory distress, hypoxemia	CXR, CT chest
ENDO	Adrenal insufficiency due to chronic steroid therapy, adrenal hematomas (rare) Acute pancreatitis (rare)	Pain in abdomen radiating to the back	Look for obesity, BMI, fat deposits in the neck and decreased range of neck movement Measure thyromental distance	US/contrast CT abdomen Serum amylase, lipase
HEME	Anemia	Fatigue, breathlessness	Pallor	Hgb level, ESR, CRP, serum total IgA, galactose deficient IgA, PT, APTT

Key References: Trnka P: Henoch-Schönlein purpura in children, *J Paediatr Child Health* 49(12):995–1003, 2013; Kurdi MS, Deva RS, Theerth KA: An interesting perioperative rendezvous with a case of Henoch-Schönlein purpura, *Anesth Essays Res* 8(3):404–406, 2014.

Perioperative Implications

Preoperative Preparation

- Assess and improve volume status if vomiting/GI bleed/renal dysfunction present.
- Correct anemia if severe GI bleed/renal failure.
- Steroid supplementation if on steroids.
- If on ACE inhibitors, skip dose 24–48 h before surgery.
- IV access may be difficult because of deep-seated veins due to steroids and purpuric rash.

Monitoring

- Routine monitoring.
- Kidney and cardiac function monitoring are important. Look for bradycardia and arrhythmias.

Airway

- Cervical joint arthritis (rarely) and obesity with fat deposition on the neck and chin due to steroid therapy can lead to limited neck extension, so a difficult airway is a possibility. Keep difficult airway cart ready.
- Avoid invasive airway access.

Technique/Induction

- No preference for any technique in general exists; go for rapid sequence induction if administering general anesthesia for emergency abdominal surgery.
- Opt for regional if neck and cervical joint movements are restricted.
- Choose anesthetic drugs depending on renal, CNS, CV status, endocrine status, and presence of hypertension.

Maintenance

- Take care of joints and pressure points. Presence of arthritis, joint effusions, skin blisters, and ulcers should be kept in mind during positioning.
- Ensure sufficient IV fluid administration.
- Choose drugs like isoflurane/sevoflurane for maintenance and atracurium for neuromuscular blockade, keeping in mind renal involvement.

Postoperative Period

- The disease can worsen. Monitor and maintain renal function. Watch for cardiac arrhythmias.
- Avoid NSAIDs for pain relief if GI hemorrhage/renal involvement are present.
- Continue ACE inhibitors that were skipped preop.

Anticipated Problems/Concerns

- Risk of tissue compression and necrosis over pressure points due to positioning, BP cuff, and endotracheal intubation.
- Varying degrees of hypoxemia due to IgA deposits and periop alveolar hemorrhages; may require postop ventilator support.
- Anemia, hypoproteinemia, fluid, and lyte imbalance may increase morbidity.
- Insufficient supplementation of steroids may lead to precipitous hypotension due to acute adrenergic crisis.
- AV block, bradycardia, and cardiac arrhythmias with need for ventricular pacing and death can occur; nevertheless cardiac involvement is extremely rare.