

Risk

- Rare: 2–5 new cases/million/y
- Average age of onset: 35–50 y
- Male = female

Perioperative Risks

- Necrotizing eosinophilic vasculitis involving respiratory (100% of pts), cardiac (30–60% of pts), neurologic, GI, renal, and integumentary systems

Worry About

- Respiratory complications from severe asthma exacerbations
- CV collapse from coronary artery vasculitis, myocarditis, or cardiac tamponade; represents the major cause of mortality
- Peripheral and central neurologic defects (mononeuritis multiplex and cerebral infarcts)
- GI vasculitis (abdominal pain and bleeding)
- Effects of the standard treatments (steroids and immunosuppressants)

Overview

- Syndrome includes (1) a history of late-onset asthma, (2) eosinophilia, and (3) systemic vasculitis in two or more organ systems.
- Most pts have generalized symptoms, but respiratory effects such as asthma and pulmonary infiltrates are a core clinical feature.
- Affects all major organ systems to varying degrees.
- Delayed diagnosis is common because the first manifestation is usually synonymous with asthma and allergic rhinitis.
- Diagnosis should be considered in pts with asthma and an increased blood eosinophil count or pulmonary infiltrates.
- Prognosis: Remission can be obtained in >80% of pts, but relapse does occur.
- 5-y survival is 80%.
- Asthmatic symptoms usually persist despite recovery from vasculitic symptoms requiring chronic steroid use.

Etiology

- Attributed to an immune reaction to inhaled allergens, but the cause is unknown.
- Possible link to leukotriene receptors antagonist use.
- Three successive phases: (1) asthma and allergic manifestations → (2) blood eosinophil and tissue eosinophil infiltration → (3) systemic phase with subsequent necrotizing vasculitis.
- Clinical features tend to divide into two phenotypes: A vasculitic type with manifestations caused by small-vessel vasculitis (purpura, mononeuritis, and renal), and an eosinophilic type, where organ damage is a result of eosinophil infiltration (pulmonary and cardiac involvement). ANCA-positive pts tend to have the vasculitic type.

Usual Treatment

- Corticosteroids +/- immunosuppressant, depending on severity of organ involvement
- Pulse steroid with long-term wean if possible
- IVIG or plasma exchange for refractory cases

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Sinusitis	Headache Nasal discharge	Facial tenderness	CBC, differential, CT scan
RESP	Asthma Eosinophilic pneumonia/infiltrates	Dyspnea Fever Cough	Wheezing Lung field consolidation	CXR, CT scan, PFT ABGs, bronchoscopy ± Bronchoalveolar lavage, biopsy
CV	Coronary artery vasculitis Endomyocarditis Pericarditis Pericardial effusion/tamponade	Chest pain Dyspnea	Tachycardia, S ₃ Pericardial rub Muffled heart sounds Elevated JVP	ECG, angiogram Cardiac MRI, angiogram ECHO
GI	GI vasculitis	Abdominal pain Diarrhea Melena/hematochezia		Endoscopy
CNS	Peripheral neuropathy/mononeuritis multiplex Cranial nerve palsy Cerebral ischemia/hemorrhage	Weakness Sensory deficits	Power assessment	EMG CT
RENAL	Focal segmental glomerulonephritis	Weight gain Foamy urine	Leg edema Hypertension	BUN, Cr Urinalysis
HEME				CBC, eosinophil count ANCA
DERM	Skin lesions		Palpable purpura Cutaneous nodules	Skin biopsy
METAB	Insulin resistance secondary to long-term steroid use	Polyuria Polydipsia		Glucometer, HbA ^{1C} Oral glucose tolerance test

Key Reference: Pagnoux C, Guilpain P, Guillevin L: Churg-Strauss syndrome, *Curr Opin Rheumatol* 19:25–32, 2007.

Perioperative Implications

Preoperative Preparation

- Assess asthma control, optimization of bronchodilators, and treatment with inhaled or oral corticosteroids.
- Rule out significant cardiac involvement +/- treatment for heart failure.
- Assess immunosuppressive and steroid-induced side effects.
- Stress-dose steroids if significant surgical stress is anticipated and adrenal suppression suspected.

Monitoring

- Arterial line if significant respiratory or cardiac compromise.
- Consider CVP, PA cath, or intraop ECHO as indicated.

Airway

- Airway hypersensitivity: avoid instrumentation if possible.

Preinduction/Induction

- Regional/neuraxial anesthesia if possible
- Induction with agents that minimize airway reactivity (propofol) or have bronchodilator properties (ketamine)
- Supraglottic airway device if possible to avoid precipitating bronchospasm
- Possible link with pseudocholinesterase deficiency, although not definite

Maintenance

- Volatile anesthetics with bronchodilator properties (avoid desflurane, which can lead to coughing and bronchospasm)
- Avoid histamine-releasing medications (e.g., morphine, atracurium)

- Ventilator parameters: Low tidal volume (6–8 mL/kg), longer expiratory time, avoidance of PEEP, and peak end-inspiratory plateau pressures <30 cm H₂O

Extubation

- Period prone to airway sensitivity and bronchospasm; consider deep extubation.
- Use neostigmine, which can cause bronchospasm, with caution.

Postoperative Period

- Monitor respiratory status given postop decreases in VC and FRC.
- Have an adequate plan for analgesia with consideration for epidural analgesia if appropriate.
- Manage steroid use.

Anticipated Problems/Concerns

- Severe bronchospasm