

# Scleroderma/ Systemic Sclerosis

Systemic sclerosis is a chronic progressive autoimmune collagen vascular disease characterized by inflammation and deposition of fibrous connective tissue in the skin and internal organs, resulting in severe multiorgan system dysfunction.

## ANESTHETIC CONSIDERATIONS:

- Difficult intubation (microstomia, ↓TMJ and neck mobility, bleeding from mucosal telangiectasias)
- Aspiration risk (esophageal dysmotility, ↓LES tone)
- Multisystem organ dysfunction
  - CVS: arrhythmias, ischemia, CHF, pulmonary HTN/cor pulmonale, pericarditis (hemodynamic instability), Raynaud's phenomenon
  - RESP: pulmonary fibrosis (hypoxemia, difficult ventilation)
  - GI: hypomotility (aspiration)
  - RENAL: renal failure, renal crisis
  - MSK: myopathy (sensitivity to NMBA)
- Hypovolemia
- Difficult vascular access
- Medications (immunosuppressants, corticosteroids, vasodilators)

## ANESTHETIC GOALS:

- Recognize and secure the difficult airway
- Take appropriate aspiration precautions
- Obtain adequate iv access
- Maintain hemodynamic stability
- Ensure adequate ventilation in setting of significant lung disease
- Avoid peripheral vasoconstriction (normovolemia, normothermia)
- Consider postoperative disposition

## HISTORY

- Palpitations, angina, syncope, dyspnea, orthopnea, PND
- Cough, dyspnea, home O<sub>2</sub>
- Epistaxis
- Dysphagia, GERD, early satiety
- Cyanosis/pallor of extremities with exposure to cold
- Neuropathies – sensory deficits, weakness
- Arthritis
- Muscle weakness
- Functional capacity, weight loss
- Medications
  - Corticosteroids, cyclosporine, cyclophosphamide, IFN, vasodilators

## PHYSICAL

- VS – HTN, tachypnea, hypoxemia
- CNS – neurologic deficits
- AW – careful airway examination; telangiectasias, microstomia, limited TMJ mobility, limited neck mobility
- CVS – iv access, intravascular volume status, evidence of Raynaud's, irregular pulse, evidence of CHF, evidence of pulmonary HTN (TR murmur, loud P2), evidence of cardiac tamponade
- RESP – respiratory distress, pulmonary crackles (ILD vs CHF)
- GI – abdominal distention
- DERM – thick fibrotic skin (back and buttocks spared)
- MSK – proximal muscle weakness, contractures

## INVESTIGATIONS

- Labs
  - CBC/D – anemia, pancytopenia (S/E of immunosuppressants)
  - Lytes, urea, Cr – renal insufficiency
  - INR/PTT – coagulopathy (vit K malabsorption)
  - U/A – proteinuria, hematuria
  - ABG – hypoxemia
- Imaging
  - CXR – CHF, aspiration pneumonia, interstitial lung disease
  - EKG – arrhythmias, pulmonary HTN, pericarditis
  - Echocardiogram – diastolic dysfunction, ↓EF, pulmonary HTN, pericardial effusions
- Other
  - PFTs (restrictive lung disease, ↓DLCO)

## OPTIMIZATION

- Consults
  - Cardiology/Pulmonary/GI/Nephro/Rheumatology as indicated
  - ICU for postop ventilation if severe cardiopulmonary impairment
- Optimize cardiopulmonary status
- Aspiration prophylaxis (sodium citrate, H2RA)
- Vitamin K replacement (iv)

- Stress dose steroids if chronic corticosteroid use

#### ANESTHETIC OPTIONS

- **Local anesthesia**
- **Regional anesthesia**
  - Document neuropathies preoperatively
  - Assess for coagulopathy
  - May be technically difficult due to thick fibrosed skin and contractures
  - Avoid techniques which impair respiratory mechanics (interscalene, high spinal)
  - Use epinephrine-free local anesthetics
  - Sympathetic blockade reduces vasospasm however risk of hypotension
  - Prolonged response to local anesthetics (↓vascular uptake of LA due to microvascular fibrosis)
- **General anesthesia**
  - Renal dosing of drugs in setting of impaired renal function
  - Increased sensitivity to myocardial depressants
  - May have increased sensitivity to respiratory depressant effects of opioids
  - Increased sensitivity to muscle relaxants (myopathy) – use short-acting agents not dependent on renal clearance

#### ANESTHETIC SETUP

- **Drugs**
  - Standard emergency drugs including vasoconstrictors, vasopressors
  - Consider need for inotropes, antiarrhythmics
- **Equipment**
  - Standard CAS monitors
  - 5 lead EKG
  - Temperature probe, forced air warmers, fluid warmer
  - Arterial line (reduced coronary vascular reserve, inaccurate NIBP readings due to skin fibrosis)
    - Radial artery catheterization contraindicated in Raynaud's (risk of hand ischemia); brachial artery catheterization may be necessary
  - +/- CVL
  - Consider TEE (passage of probe may be difficult if esophageal strictures) or PA catheter
  - Difficult airway cart and fiberoptic bronchoscope
  - ?ICU ventilator

#### MANAGEMENT OF ANESTHESIA

- **Induction**
  - Difficult iv access; may require central line
  - Preoxygenation
  - Consider awake tracheostomy or awake intubation
    - RSI if asleep intubation (aspiration risk)
  - Avoid airway trauma during intubation and avoid nasal intubation (bleeding from telangiectasias)
  - Monitor for hypotension during induction (intravascular volume contraction with HTN, vasomotor instability, myocardial dysfunction)
- **Maintenance**
  - Protect eyes to prevent corneal abrasions
  - Careful positioning (contractures)
  - May require high airway pressures to ensure adequate ventilation (poor pulmonary compliance)
  - Avoid factors which increase PVR (hypoxemia, hypercarbia, acidosis, N<sub>2</sub>O)
  - Minimize peripheral vasoconstriction (normovolemia, normothermia – room temperature >21°C, warm iv fluids, forced air warmer)
- **Emergence**
  - Consider need for postoperative ventilatory support if severe pulmonary disease
  - Ensure complete reversal of neuromuscular blockade and return of protective airway reflexes
  - PONV prophylaxis

#### DISPOSITION & MONITORING

- Consider postop ICU for ventilatory support if severe pulmonary disease

#### COMPLICATIONS

- Difficult/failed intubation
- Aspiration
- Hypoxemia (restrictive lung disease, pulmonary HTN)
- Cardiogenic shock

#### OBSTETRICS

- Pregnancy accelerates progression of scleroderma in 50% of patients
- Increased incidence spontaneous abortion, PTL, perinatal mortality
- Possible placental dysfunction
  - Nitric oxide donors and heparin may be useful
- Avoid ACE-I (teratogenic) *unless* overt renal crisis

#### PATHOPHYSIOLOGY

- Epidemiology
  - Typical age of onset: 20-40 yrs
  - F:M = 5:1

- Etiology
  - Unknown
  - Characteristics of collagen vascular disease and autoimmune disease
- Pathophysiology
  - Fibroblasts produce excess collagen → microvascular obliteration → tissue and organ fibrosis
  - Endothelial damage → inflammation and immune cell activation → ↑T cell and B cell activity
    - ↑Endothelin production → vasospasm
    - ↑Capillary permeability → leakage of serum proteins into interstitium → edema and lymphatic obstruction
  - Autoantibodies produced against nuclear and centromere structures
  - Some clinical similarities to GVHD
- Classification
  - Localized
    - Only skin affected
  - Limited cutaneous systemic sclerosis (aka CREST syndrome)
    - Involves skin of face and upper extremities + pulmonary and GI involvement
      - Calcinosis, Raynaud's, Esophageal dysmotility, Sclerodactyly, Telangiectasias
      - Slowly progressive
  - Diffuse cutaneous systemic sclerosis
    - Generalized skin involvement (back and buttocks spared) + multiple end-organ damage
    - More rapidly progressive
- Manifestations
  - Most common presentation
    - Triad of skin thickening, nonpitting edema, Raynaud's phenomenon
  - CNS
    - Neuropathy – CN and PN (nerve compression by thickened connective tissue)
      - Eg: trigeminal neuralgia
  - H+N
    - Keratoconjunctiva sicca (↓ tear production) → risk of corneal abrasions
    - Xerostomia → poor dentition
    - Microstomia → difficult intubation
    - Decreased TMJ and neck mobility → difficult intubation
    - Oral and nasal telangiectasias → friable mucosa
  - CVS
    - Arrhythmias (sclerosis of conduction system)
    - Ischemic heart disease (coronary artery sclerosis)
    - Pulmonary HTN → cor pulmonale (pulmonary artery fibrosis, interstitial lung disease)
    - CHF (myocardial fibrosis in 70-80%; muscle replaced with fibrous tissue)
    - Pericarditis → pericardial effusion → tamponade
    - Raynaud's phenomenon (85%; vasospasm of small arteries) → cyclic pallor and cyanosis of digits in response to cold or emotion
      - Stellate ganglion block may worsen contralateral Raynaud's (but relieves pain from ipsilateral disease)
      - β-blockers relatively contraindicated
  - RESP
    - Diffuse interstitial pulmonary fibrosis (80-90%) → hypoxemia, pulmonary HTN, poor lung compliance, ↑ risk postoperative pulmonary complications
    - Pleuritis
    - Aspiration pneumonitis
  - GI
    - Dysphagia (esophageal hypomotility 2° to muscle atrophy, mucosal fibrosis) → aspiration
    - GERD (↓ LES tone) → esophagitis, aspiration
    - Intestinal pseudoobstruction (intestinal hypomotility) → ileus, constipation, overflow diarrhea
    - Small bowel bacterial overgrowth (intestinal hypomotility) → malabsorption → vitamin K deficiency → hypocoagulability
  - Renal
    - Renal artery stenosis (arteriolar intimal proliferation) → ↓RBF, HTN, CRF
    - Scleroderma renal crisis (HTN, retinopathy, rapid deterioration in renal function)
      - May be precipitated by corticosteroids
  - Derm
    - Skin thickened → atrophy/fibrosis → taut skin → limited joint mobility, flexion contractures, sclerodactyly, difficult iv access
    - Diffuse nonpitting edema
  - MSK
    - Myopathy → proximal muscle weakness, ↑plasma CK
    - Mild inflammatory arthritis (symmetric, small joints)
    - AVN femoral head
- Diagnosis
  - Autoantibodies (levels may correlate with disease activity)
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- Treatment
  - Modification of disease process
    - Immunosuppressants

- Corticosteroids – ↑ glucose, HTN, weight gain, adrenal insufficiency, AVN, etc
    - Cyclophosphamide – WBC, hemorrhagic cystitis, inhibits pseudochoolinesterase
  - Statins to reduce inflammation
  - Endothelin antagonists to inhibit effect of endothelin
- Cardiac dysfunction
  - Vasodilators (CCB, ACE-I, prostacyclin) for cardiac dysfunction, pulmonary HTN, Raynaud's
- GERD
  - PPIs
- Intestinal hypomotility
  - Somatostatin analogues beneficial (octreotide)
    - Prokinetic agents ineffective
  - Antibiotics for small bowel overgrowth
- Renal crisis
  - ACE-inhibitors
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- Outcomes
  - Renal failure and malignant HTN are most common causes of death

#### REFERENCES

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