Systemic Lupus Erythematosus

SLE is a multisystem chronic inflammatory disease characterized by antinuclear antibody production, potentially affecting nearly every organ system; of particular concern are changes to the cardio-respiratory system and hematologic changes

ANESTHETIC CONSIDERATIONS:

- Potential **difficult airway**
  - Deforming arthritis
  - Cricoarytenoid arthritis
- Systemic Complications of SLE
  - CNS - seizure, stroke, mood, organic disease
  - Cardiac - pericarditis, endocarditis, **CAD**, AI, MR, HTN / pHTN
  - Respiratory - infiltrates, infection, **restrictive lung disease**
  - Hematologic - anemia, **thrombocytopenia, abnormal bleeding / clotting**
  - Nephritis
- Complications related to medical management
  - **Steroids**
  - Immunosuppressants / antimalarials
  - NSAIDs
  - Anticoagulants, ASA
- Potential for exacerbation of SLE with surgery, stress, infection and pregnancy

ANESTHETIC GOALS:

- Preoperative optimization of multisystem disease.
- Perioperative continuation of immunosuppressant medications.
- Avoid drugs that can cause relapse (procainamide, phenytoin, penicillin, hydralazine)
- Avoid precipitants of pulmonary hypertension: acidosis, hypoxemia, hypercarbia, SNS stimulation
- Stress dose steroids

HISTORY

- Medications – steroids, NSAIDs, ASA, heparin, methotrexate, antimalarials
- Disease quiescence, progression, symptoms, organ systems involved
- Functional capacity for pulmonary and cardiovascular reserve
- Renal insufficiency
- Abnormal bleeding history, DVT / PE, strokes, spontaneous hemorrhage, pregnancy losses

PHYSICAL

- **HEENT**
  - Focused airway examination
- **CNS**
  - Peripheral neuropathies / neurological deficit documentation
- **CVS**
  - Murmurs, rubs, pulmonary edema
- **RESP**
  - Infections, chest wall excursion / paroxysmal breathing (diaphragmatic dysfunction and “shrinking lung syndrome”)

INVESTIGATIONS

- **Labs**
  - CBC, lytes, BUN, Cr, coags, X-match if indicated for procedure (may have difficult antibody screen)
  - Factor assays for deficiency in presence of prolonged PTT (most common = factor VIII)
  - Urinalysis (protein)
  - Autoantibodies – ANA, antiphospholipid, lupus anticoagulant, anticardiolipin
- **Imaging**
  - ECG, echo if indicated, cardiac investigations as indicated by functional status
  - CXR, PFTs if indicated by history
- **Special**
  - Hematology, rheumatology consultation

OPTIMIZATION

- Consultation with hematologist, rheumatologist, obstetrician and neonatologist (for the parturient)
- Management of current complications – pulmonary infections, drainage of effusions
- Antibiotic prophylaxis for valvular lesions or endocarditis
- Steroid supplementation for the steroid dependent

ANESTHETIC OPTIONS

- Local, regional, neuraxial, general
- Consider type of surgery, presence and severity of systemic disease
- Prolonged PTT that is associated with antiphospholipid antibody is not a contraindication to neuraxial, but factor deficiency with prolonged PTT is a contraindication
- Platelet count and clinical indications of platelet dysfunction may contraindicate neuraxial techniques

ANESTHETIC SETUP

- **Drugs**
  - Standard emergency drugs
- **Equipment**
  - CAS + 5 lead
  - Arterial line if significant cardiorespiratory disease
  - Other monitors as dictated by procedure (CVC, Foley, temperature, warmers)
  - Consider TEA for post-op analgesia for appropriate procedures with lung disease secondary to SLE

MANAGEMENT OF ANESTHESIA

- **Induction**
  - RA: contraindicated of coagulopathy is present
  - GA:
    - Awake FOI if anticipated difficult airway (C-spine involvement is rare)
    - Hypersensitivity to NDMR and DMR
    - If on cyclophosphamide – prolonged response to SCh and mivacurium due to inhibition of plasma cholinesterase
- **Maintenance**
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- **Emergence**
  - Fully reversed, AWaC

DISPOSITION & MONITORING

- Dictated by severity of disease, possibly high acuity unit
- Consider post-op ventilation for those with significant restrictive lung disease
- 30% recurrent laryngeal nerve involvement – consider as cause of postoperative respiratory compromise
- Prone to infection and pulmonary complications
- Treat laryngeal edema / stridor with IV steroids

COMPLICATIONS

- CHF, arrhythmia
- Respiratory insufficiency
- Renal insufficiency – volume overload

PATHOPHYSIOLOGY

- SLE has a 10:1 female preponderance, occurring most commonly in the 2^{nd}-4^{th} decade
- Relapsing / remitting course varying from mild to severe
- Constitutional symptoms occur in 50-100% of patients, fatigue being the most common and often most debilitating
- **Neurologic complications**
  - Cognitive deficits, organic brain syndromes, delirium, psychosis, seizures, headaches, depression, anxiety and mania
  - Can be secondary to the SLE disease process itself, secondary to medications, or related to thromboembolic disease in the presence of antiphospholipid antibodies
- **Cardiovascular**
  - Pericarditis, verrucous endocarditis (non-infectious – libman sacks endocarditis) usually clinically silent, but produces valvular insufficiency and a source of emboli (mitral and aortic valves)
  - Accelerated atherosclerosis
  - HTN
- **Respiratory**
  - Pleurisy, effusions, interstitial lung disease, pulmonary hypertension and alveolar hemorrhage
  - Increased risk of DVT / PE with antiphospholipid antibodies
  - In general, PFTs in SLE will show restrictive pattern (sometimes in the absence of significant disease = "shrinking lung syndrome")
- **Hematologic**
  - Leukopenia, thrombocytopenia and anemia common
  - Some patients will have antiphospholipid antibodies predisposing to thromboembolic disease and adverse pregnancy outcomes
- **Renal disease**
  - Approximately 50%, most of these being sub-clinical
  - Several forms of glomerulonephritis: proteinurea, hypertension, nephritic syndrome, renal insufficiency
- **GI effects**
  - Most commonly a side effect of medications (NSAID and steroid gastritis, ulcers)
  - SLE vasculitis can result in pancreatitis, peritonitis and colitis
- **Musculocutaneous**
  - Arthritis: joint symptoms in > 90%, often the earliest manifestation, usually migratory and asymmetrical arthritis
  - Avascular necrosis, most often of head or condyle of femur
  - Myopathy, proximal skeletal muscle weakness, increased CK, tendonitis
  - Mucocutaneous – skin lesions – most common is the butterfly malar rash
    - Painless oral or nasal ulcers also occur
    - Typical rash – discoid butterfly rash or malar rash, maculopapular rash, exacerbated by sun, alopecia
- **Ophthalmologic**
  - Keratoconjunctivitis sicca
- **SLE in Pregnancy** – exacerbations in maternal disease occur in 30-50% of pregnancies
  - Pregnancy does not alter the long term course of the disease
  - Spontaneous abortion rate 2 x the frequency of population, fetal death in ⅓ of SLE pregnancies
  - Potential for neonatal lupus – anti-Ro Ab crosses placenta causing transient symptoms, but cardiac conduction defects are permanent (congenital heart block)

- Other complications associated with pregnancy and SLE include: increased incidence of PIH, thrombocytopenia

- **Drug-induced lupus:**
  - May be caused by procainamide, quinidine, hydralazine, methyldopa, captopril, enalapril, clonidine, isoniazid, or minocycline
  - The clinical manifestations are generally mild and include arthralgia, fever, anemia, and leukopenia
  - These effects typically resolve within 4 weeks of discontinuation of the drug, although some patients have significant morbidity
- **Management:**
  - Immunosuppressants (corticosteroids) or cytotoxic drugs (cyclophosphamide, azathioprine, cyclosporine)
  - Bone marrow transplantation for the treatment of SLE is under investigation
  - NSAIDs are the first line treatment for arthritis; however, these drugs can cause renal dysfunction
  - The antimalarials, hydroxychloroquine and chloroquine, are administered for the treatment of arthritis if treatment with NSAIDs is unsatisfactory

REFERENCES

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- uptodate.com – Systemic Lupus Erythematosus
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