Pulmonary Manifestations of SLE
Cheema GS, Quismorio FP. Interstitial lung disease in systemic lupus erythematosus. 
Murray and Nadel: Textbook on Respiratory Medicine, 3rd ed.
Primer on the Rheumatic Diseases, 12th ed; Chapter 17: SLE.
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**Key Points:**
- Pleurisy is the most common form of pulmonary disease in SLE
- Both acute lupus pneumonitis and diffuse alveolar hemorrhage are fairly uncommon but have a high mortality
- Chronic lupus pneumonitis/lupus-associated ILD resembles IPF clinically
- Don’t forget to exclude infection, infection, infection

1. Pleurisy: May or may not be associated with pleural rub and/or effusion. If effusion is present, it will usually be an exudate with low complement, mildly low glucose, and high LDH but low total protein.
   - Prevalence in one series: 44% with pleurisy, 20% with effusion.

2. Acute lupus pneumonitis: Fairly uncommon (1-12% of patients).
   - Presents with fever, cough +/- hemoptysis, SOB, bibasilar fluffy infiltrates
   - ?Association with anti-dsDNA, anti-Ro/SSA antibodies
   - Chest CT usually demonstrates ground glass infiltrates
   - Pathologic findings are mixed but frequently include diffuse alveolar damage and alveolitis
   - Mortality 50%; survivors left with restrictive lung disease

3. Chronic lupus pneumonitis: Sometimes follows an acute episode; prevalence < 5%
   - Clinically similar to IPF with bibasilar crackles and infiltrates
   - PFTs with restrictive pattern
   - Chest CT may demonstrate a ground glass appearance (consistent with biopsy showing cellular inflammation) or a reticulonodular appearance (consistent with biopsy showing fibrotic pattern); this differentiation will help determine treatment

4. Pulmonary HTN: 12-28% of patients with SLE
   - Clinical presentation as with other pulmonary HTN patients
   - Associated with Raynaud’s phenomenon, elevated RF titer, anti-RNP, and anti-phospholipid antibodies

5. “Shrinking lung” syndrome: Essentially restrictive lung disease and pleuritic chest pain in the absence of any documented pulmonary disease on CT
   - Etiology unclear, although diaphragms usually elevated so ?diaphragmatic weakness

6. Diffuse alveolar hemorrhage: Fairly uncommon; in one series, 3.7% of patients
   - Pathology may demonstrate either capillaritis or “bland hemorrhage”

7. Pulmonary embolism: Frequently associated with anti-phospholipid antibody syndrome

8. Less common: BOOP, other complications of APLS

9. Don’t forget about infection (including PCP and other unusual pathogens in patients on high dose steroids), cardiogenic pulmonary edema, costochondritis