Systemic Lupus Erythematosus

- A multi-system autoimmune disease
- Etiology unknown
- Most common in women, young to middle–aged
- Highly variable clinical presentation in individual patients

SLE, Systems Involved-(1)

- Skin: photosensitive erythematous eruption
- Joints: inflammatory arthritis, usually symmetrical without joint destruction
- Kidneys: Glomerulonephritis
- Cardiopulmonary: Pleurisy, pericarditis, pneumonia and pulmonary hemorrhage, myocarditis and coronary artery disease

SLE, systems involved-(2)

- Central nervous system: Seizures, psychiatric symptoms, cerebrovascular accidents
- Blood: Anemia, often hemolytic. Leukopenia. Thrombocytopenia
- Fever

SLE: Anti-Nuclear Antibodies

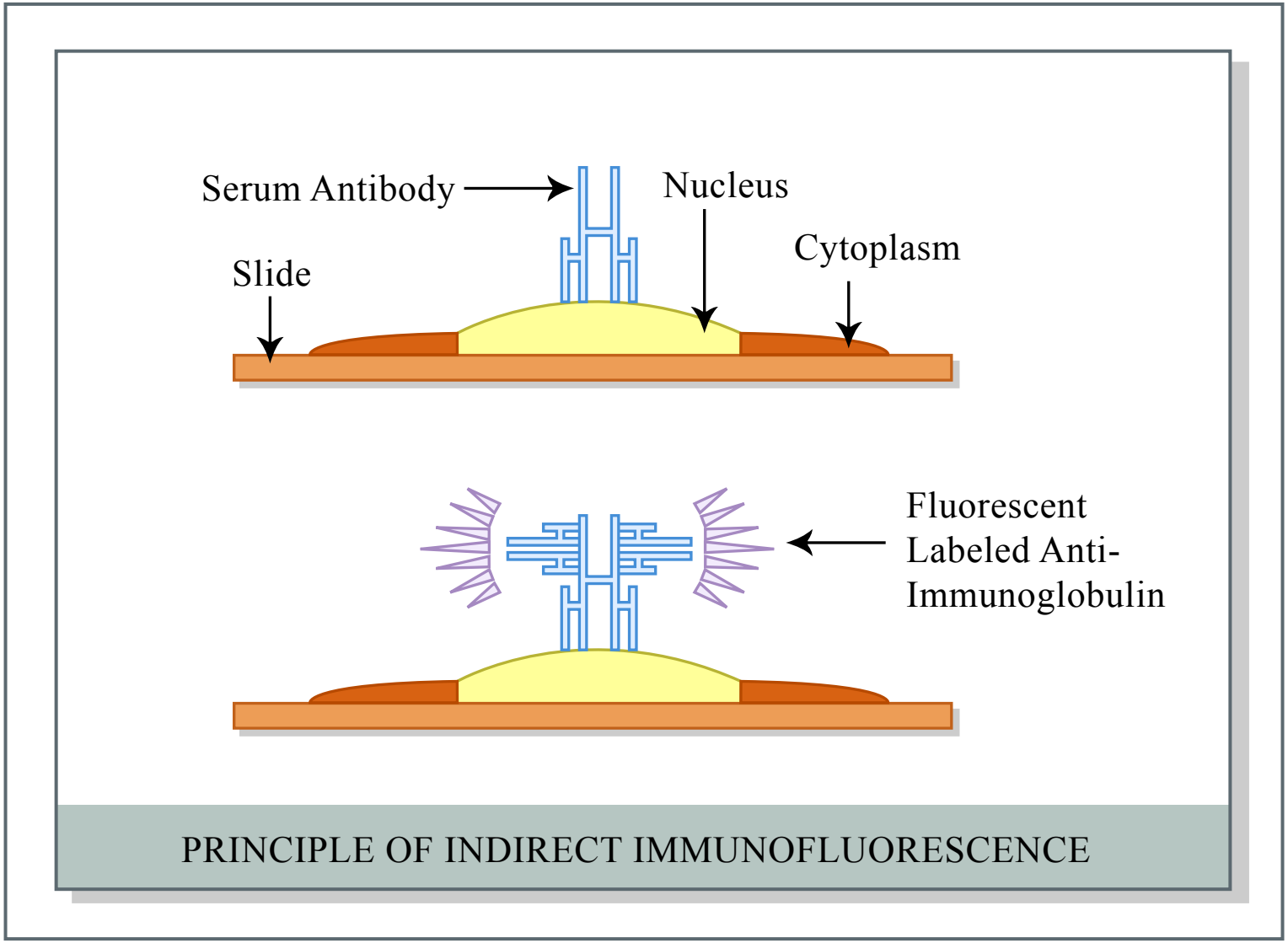

Figure by MIT OCW.

SLE: Prognosis and Treatment

- Prognosis varies from mild to severe or fatal.
- Treatment is non-specific, and is not curative.
  - NSAIDs
  - Hydroxychloroquine
  - Glucocorticoids
  - Cytotoxic, immunosuppressive drugs

Diffuse Systemic Sclerosis (Scleroderma)

- A chronic, progressive inflammatory auto-immune disease leading to fibrosis in several organ systems and in the vasculature
- Etiology unknown
- No specific treatment; only palliative measures with limited efficacy.

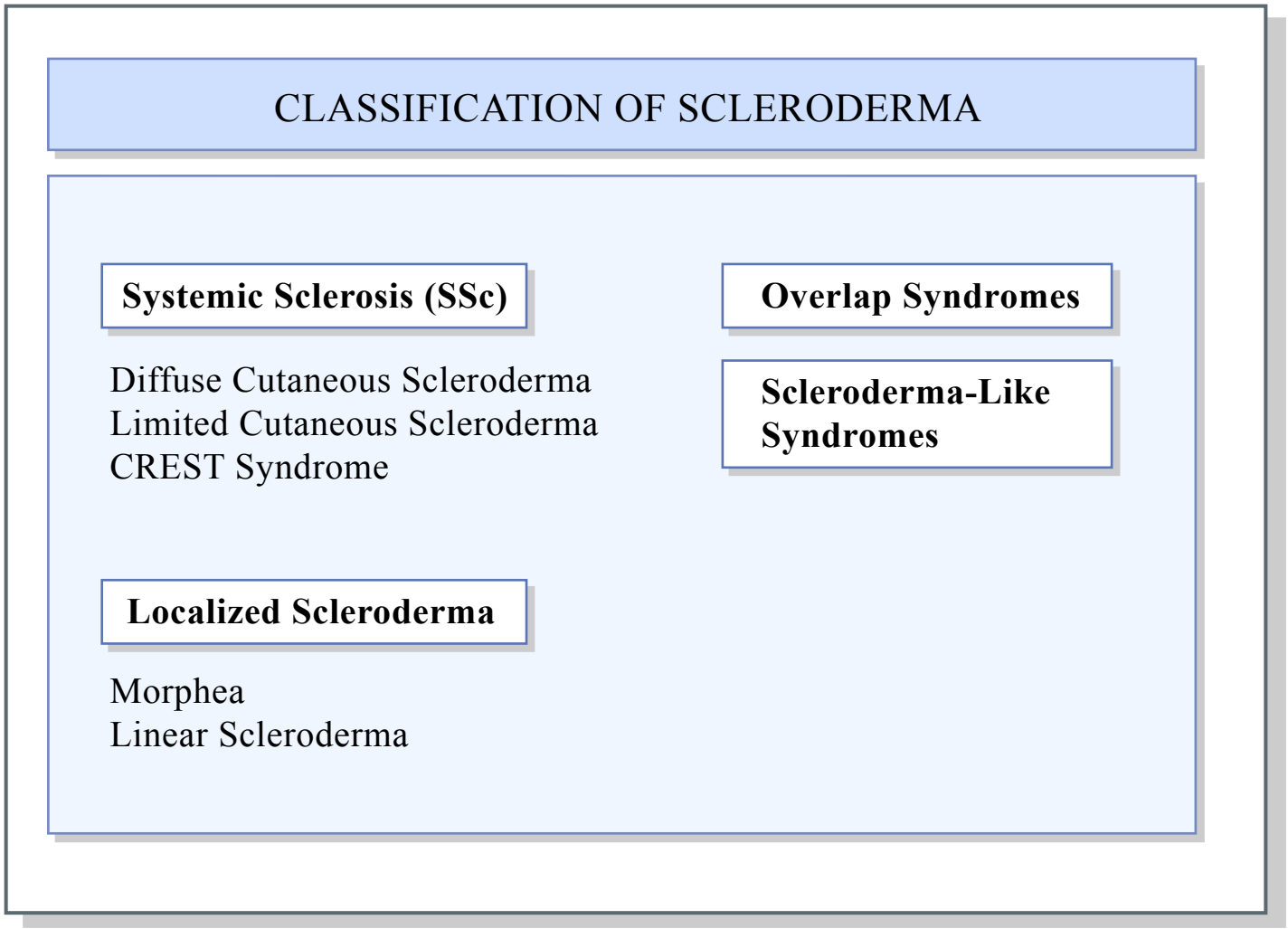

Figure by MIT OCW.

Idiopathic Inflammatory Myopathy

- Inflammatory myopathy of unknown etiology
- Probably has an autoimmune pathogenesis
- Usually a chronic progressive disease
- Causes proximal skeletal muscle weakness
- This entity and other diseases of muscle will be discussed by Drs. Brown and Johns

POLYMYOSITIS: CLASSIFICATION

- Adult Polymyositis
- Adult Dermatomyositis
- Inflammatory Myositis Associated with Cancer
- Childhood Dermatomyositis or Polymyositis
- Myositis Associated with Connective Tissue Disease

Figure by MIT OCW.

Idiopathic Inflammatory Myositis Diagnosis

- Elevation of serum levels of enzymes intrinsic to skeletal muscle; creatine phosphokinase is the most sensitive and specific. Transaminases also are elevated.
- Myopathic changes on EMG
- Abnormal muscle biopsy
- Evidence of inflammation on MRI

Idiopathic Inflammatory Myositis Treatment

- Glucocorticoids: Relatively high doses required. Toxicity is frequent.
- Other immunosuppressive agents: methotrexate, azathioprine
- Physical therapy
- Search for underlying malignancy where appropriate

Sjogren's Syndrome (Keratoconjunctivitis sicca)

- An inflammatory auto-immune disease involving the salivary and lacrymal glands, sometimes other exocrine glands.
- Etiology unknown
- Causes dryness of the eyes and mouth.
- May be associated with other rheumatic diseases, such as rheumatoid arthritis and SLE.
- Treatment is palliative

Vasculitis

- A bewildering array of clinical syndromes with the common feature of necrotizing inflammation of blood vessels.
- The etiology is often unknown, but some infections, e.g., hepatitis C, can cause vasculitis through immune complex deposition

Vasculitis may be classified on the basis of the size of the arteries involved

VASCULITIS SYNDROMES	
Vasculitis Syndrome	**Vessel Involved**
Polyarteritis Nodosa	Small, Medium Arteries
Churg-Strauss (Allergic Granulomatosis and Angiitis)	Small, Medium Arteries
Hypersensitivity Vasculitis	Arterioles, Venules, Capillaries, Rarely Small Arteries

Figure by MIT OCW.

VASCULITIS SYNDROMES (Cont.)

Vasculitis Syndrome	Vessel Involved
Henoch-Schönlein Purpura	Venules, Arterioles, Capillaries
Takayasu's Arteritis	Medium, Large Arteries
Temporal Arteritis	Medium, Large Arteries
Wegener's Granulomatosis	Small Arteries, Veins, Medium Arteries

Figure by MIT OCW.

Vasculitis: Diagnosis

- Recognition of clinical syndromes caused by vasculitis
- Elevated levels of acute phase reactants
- Positive test for anti-neutrophil cytoplasmic antibodies (ANCA), are present in some syndromes
- Biopsies and radiographic studies

Vasculitis: Treatment

- Immunosuppressive therapy with glucocorticoids and cytotoxic or antimetabolic drugs
- Prognosis is guarded, but most syndromes are treated effectively, although cures are not always achieved