

# **Clinical Aspects of Peripheral Nerve and Muscle Disease**

**Roy Weller**

**Clinical Neurosciences**

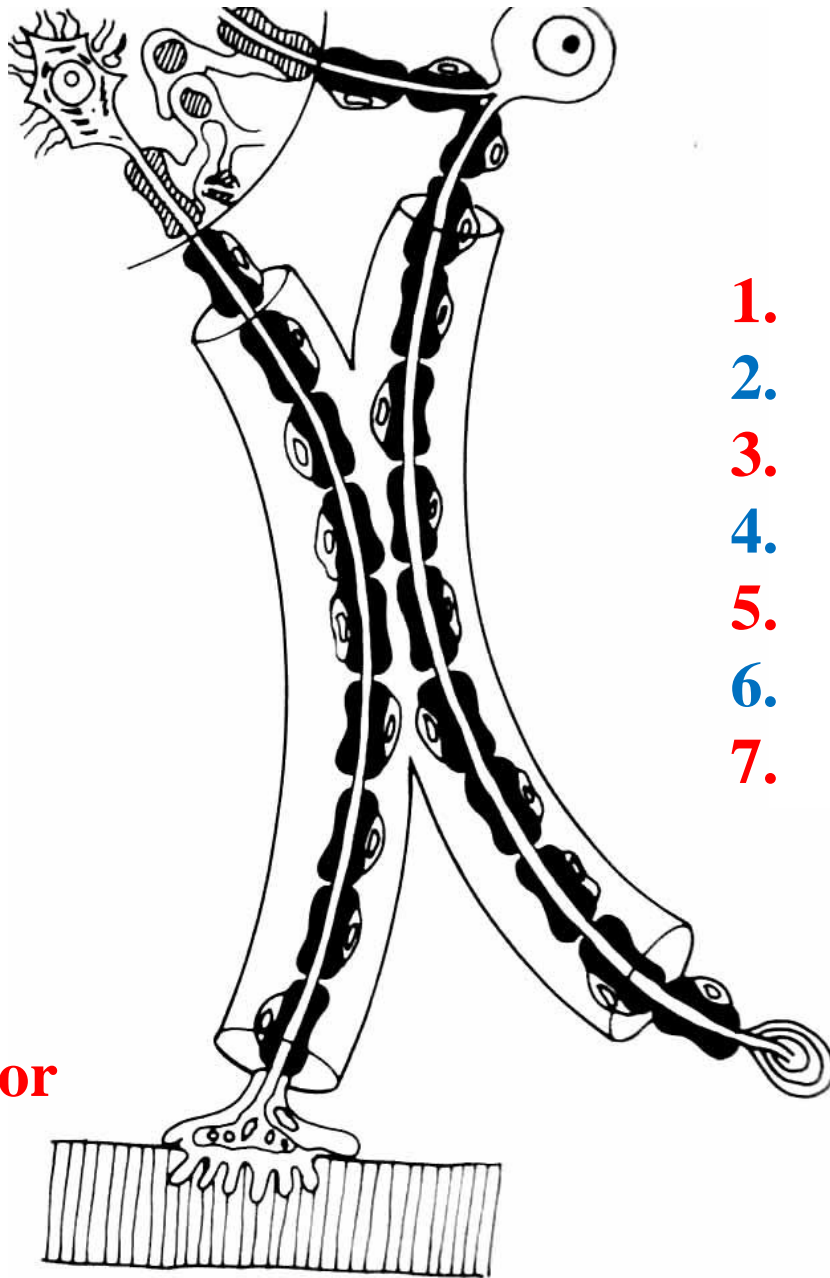
**University of Southampton School of Medicine**

# Normal Nerves

1. **Anterior Horn Cell**
2. **Dorsal root ganglion cell**
3. **Motor Peripheral Nerve**
4. **Sensory Peripheral Nerve**
5. **Neuromuscular Junction**
6. **Sensory ending**
7. **Muscle**

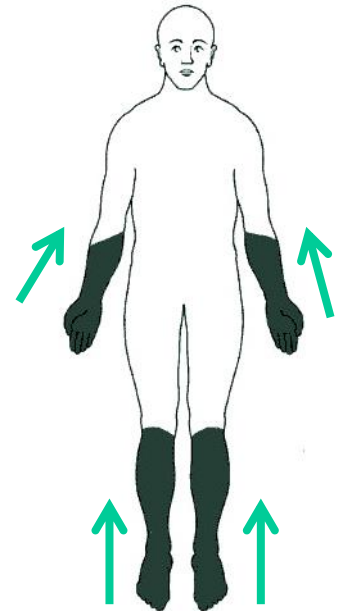
**Motor**

**Sensory**



# Peripheral Neuropathy

- **Mononeuropathy**
- **Mononeuropathy Multiplex**
- **Polyneuropathy**
- **Distal distribution of Sensory and motor signs and symptoms**

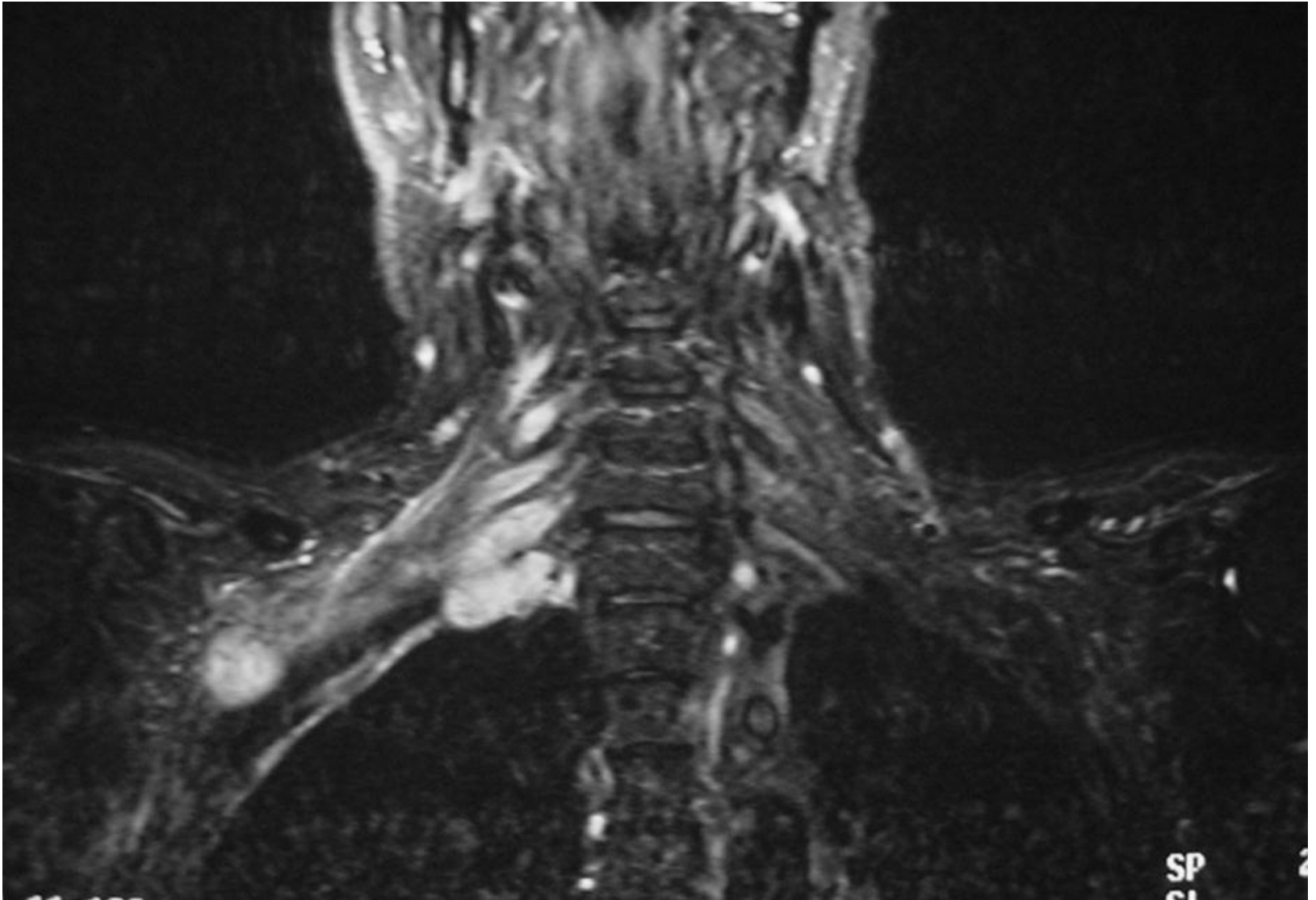


# Peripheral Neuropathy

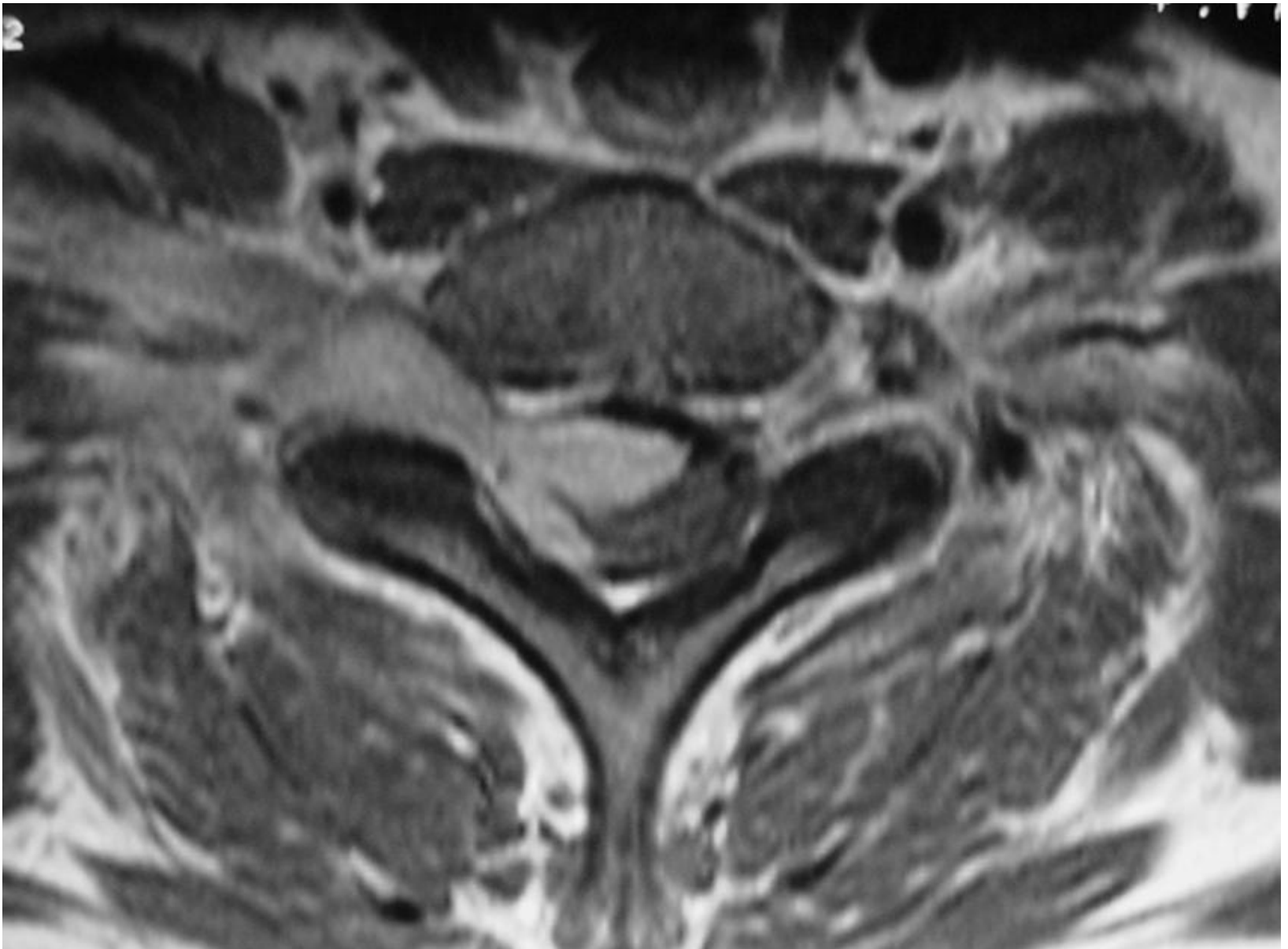
- 1. Sensory:**
  - a. Paraesthesia (tingling, pins & needles, prickling, burning...)**
  - b. Numbness (Hypoaesthesia)**
  - c. Abnormal degrees and types of sensation**
  
- 2. Motor:**
  - a. Weakness**
  - b. Wasting**
  - c. Muscle twitching (fasciculations)**

# Peripheral Neuropathy Investigations

- **Blood tests: Part of the general investigation**
- **CSF:**
  - **Protein raised in inflammatory conditions**
- **Radiology (CXR, MR Imaging)**
- **Nerve Conduction Studies + EMG**
  - **Axonal degeneration Vs Demyelinating**



**MRI of cervical Nerve root tumours**



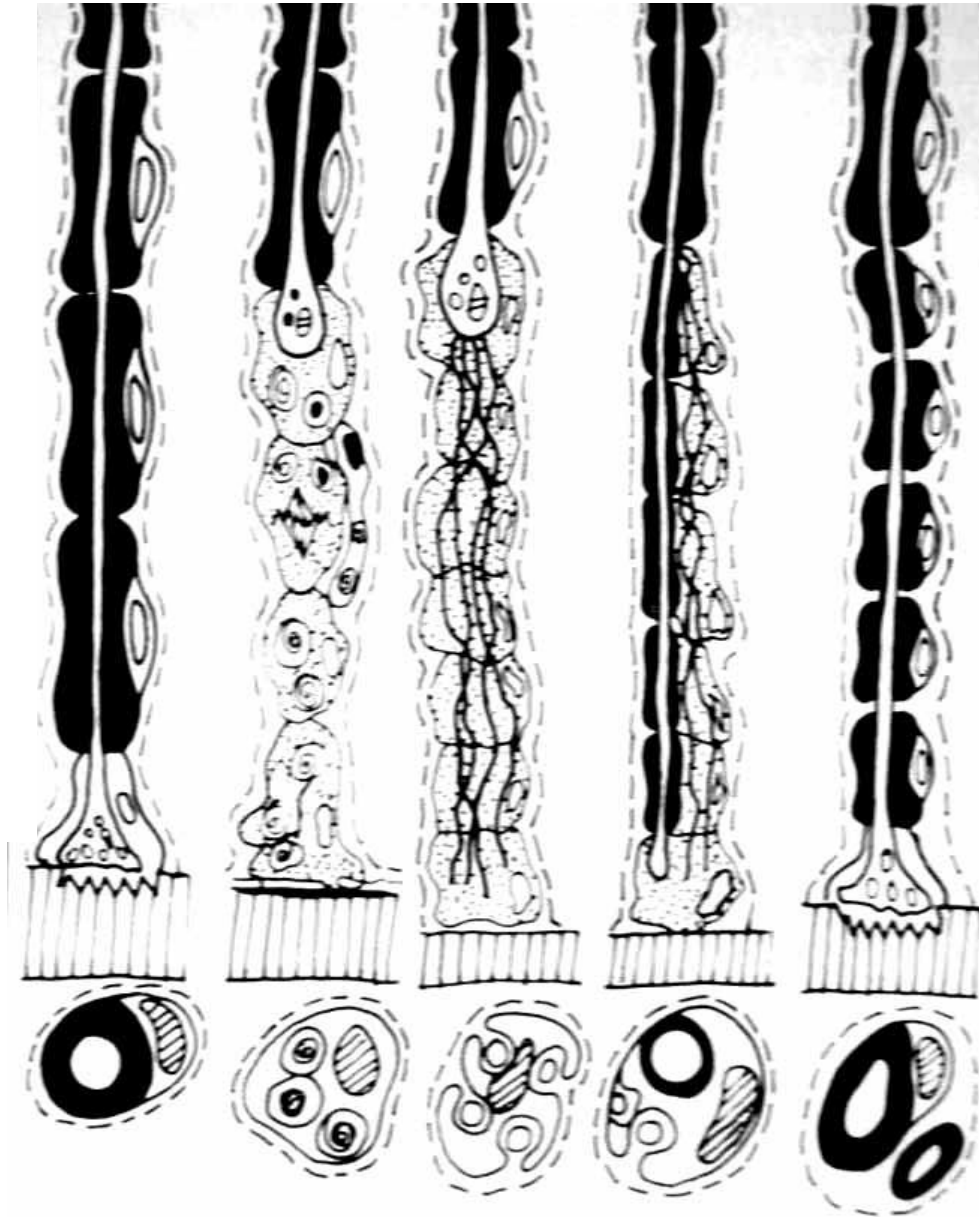
**MRI of cervical Nerve root tumour**

# Peripheral Neuropathy Investigations

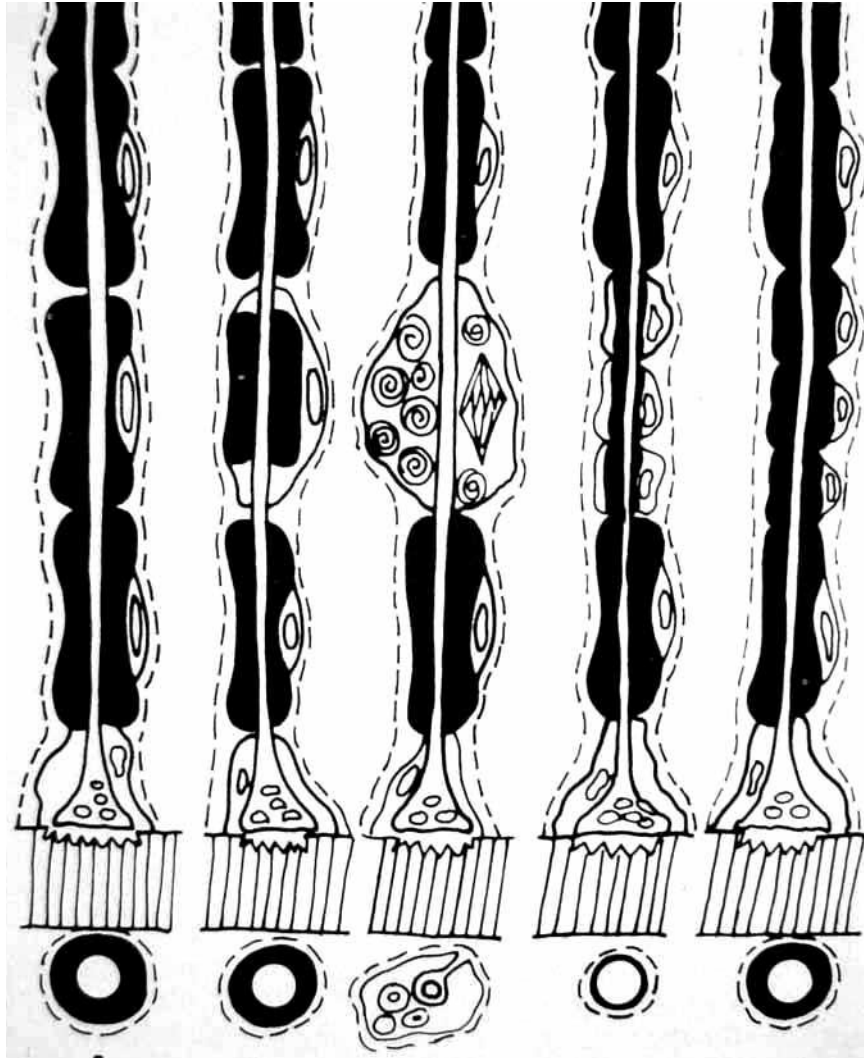
- **Nerve Biopsy**
  - **Axonal degeneration**
  - **Segmental Demyelination**
  - **Inflammatory changes**
  - **Others (Amyloid, paraprotein,..)**



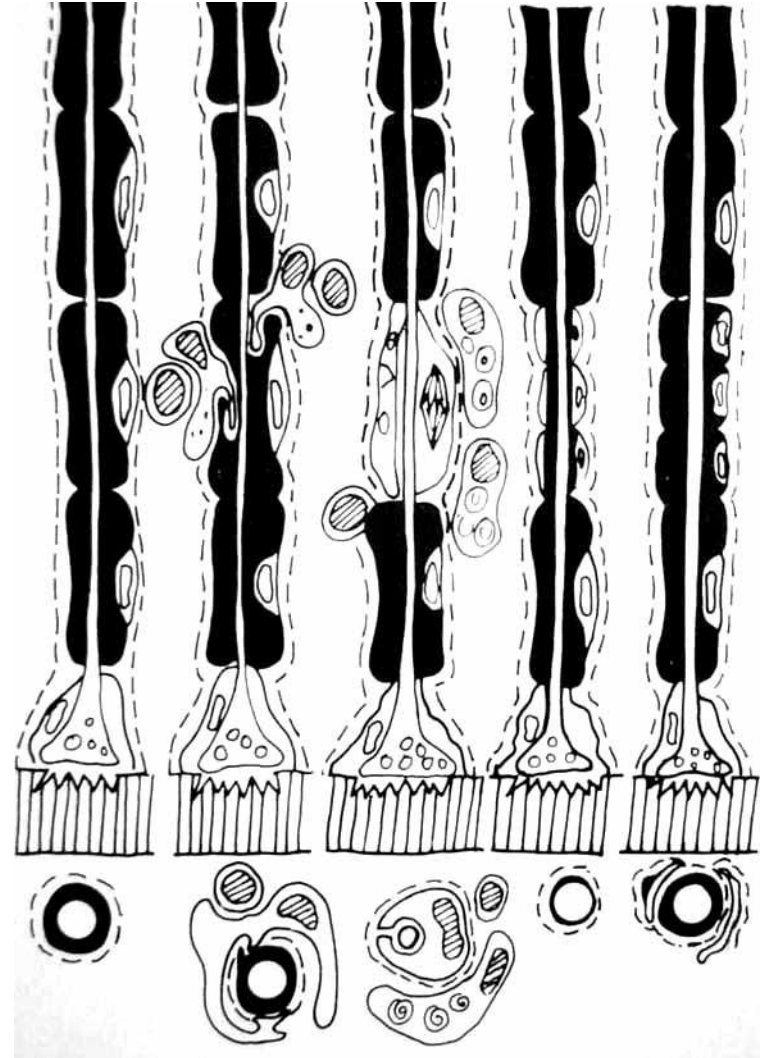
# Axonal Degeneration and Regeneration



# Segmental Degeneration

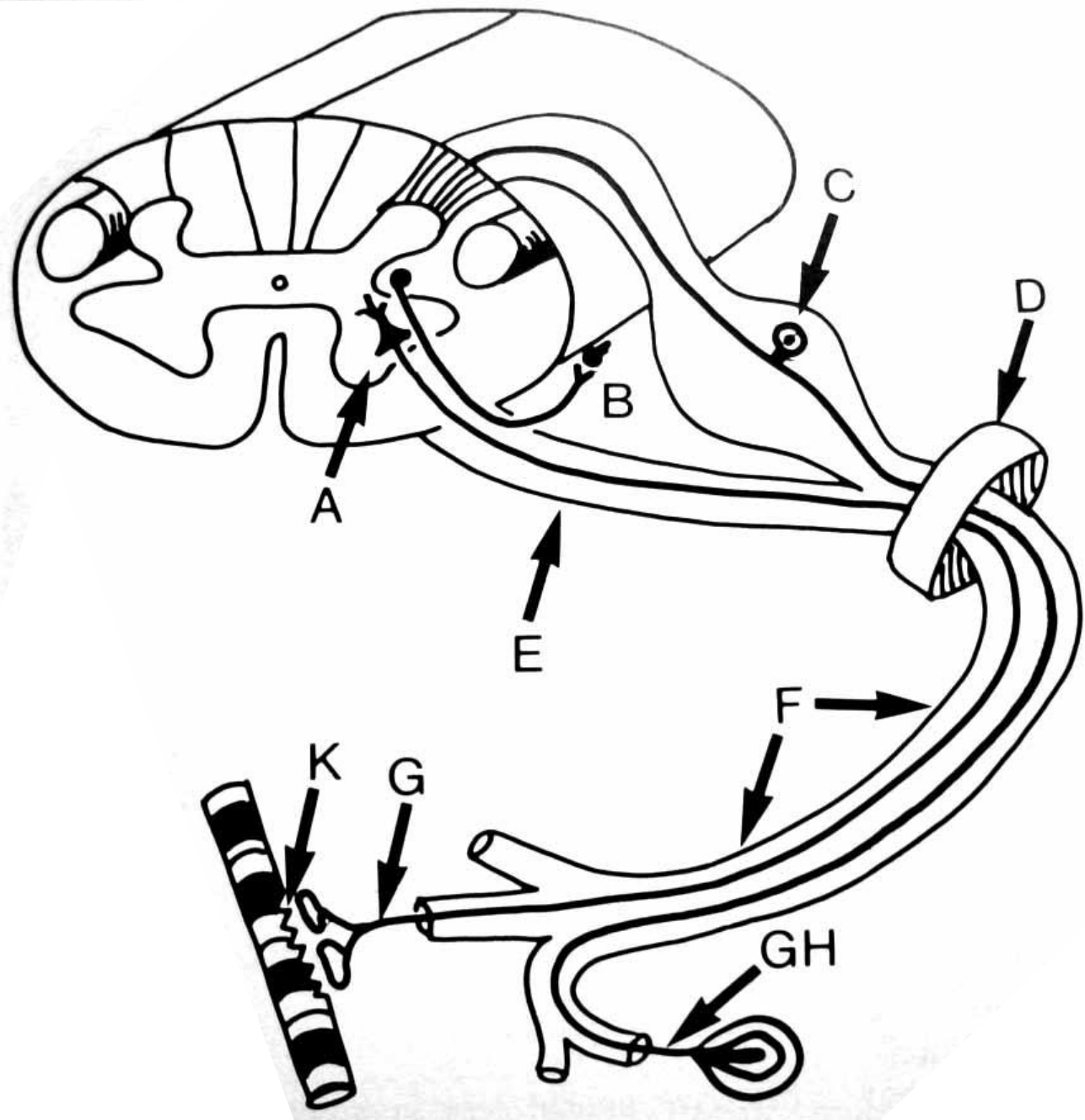


# Autoimmune Segmental Degeneration



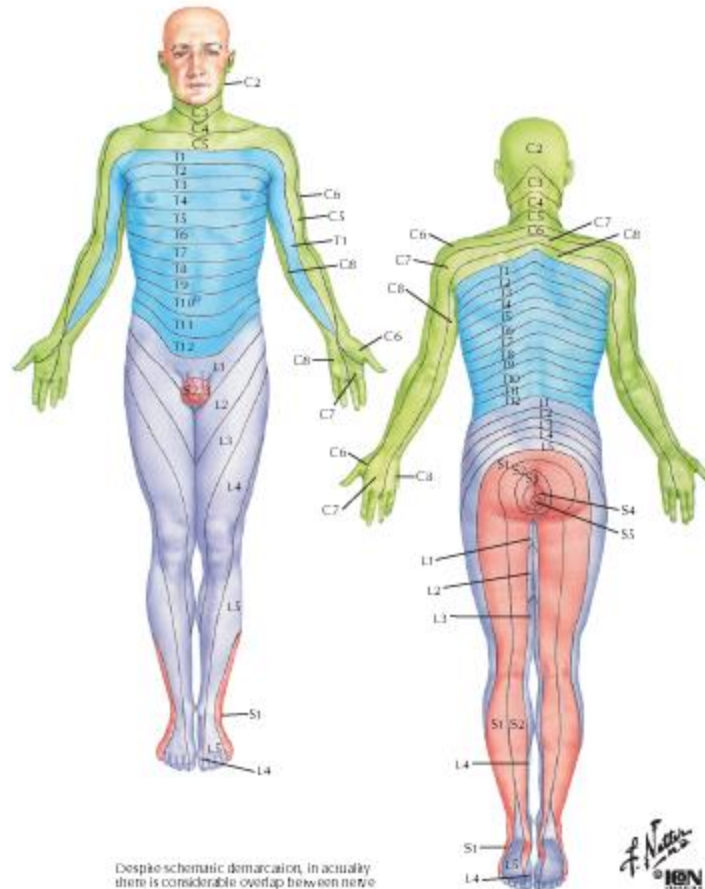
# **Clinical Approach to Peripheral Neuropathy**

- **Which systems are involved?**
- **What is the distribution of weakness?**
- **What is the nature of the sensory involvement?**
- **What is the temporal evolution? Acute, Subacute, Chronic, relapsing,..**
- **Is there evidence of heredity?**



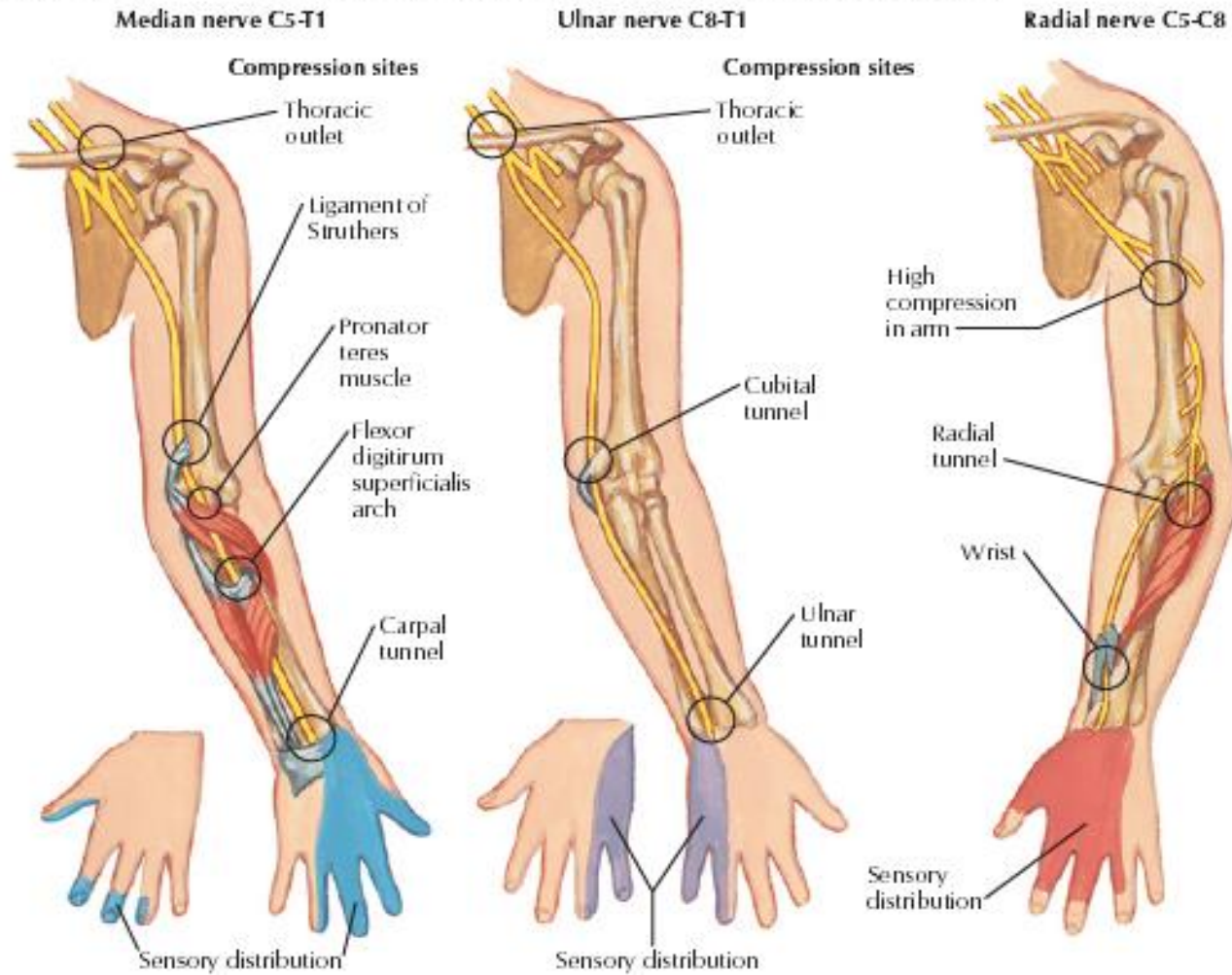
# Where is the pathology?

## Dermatomes

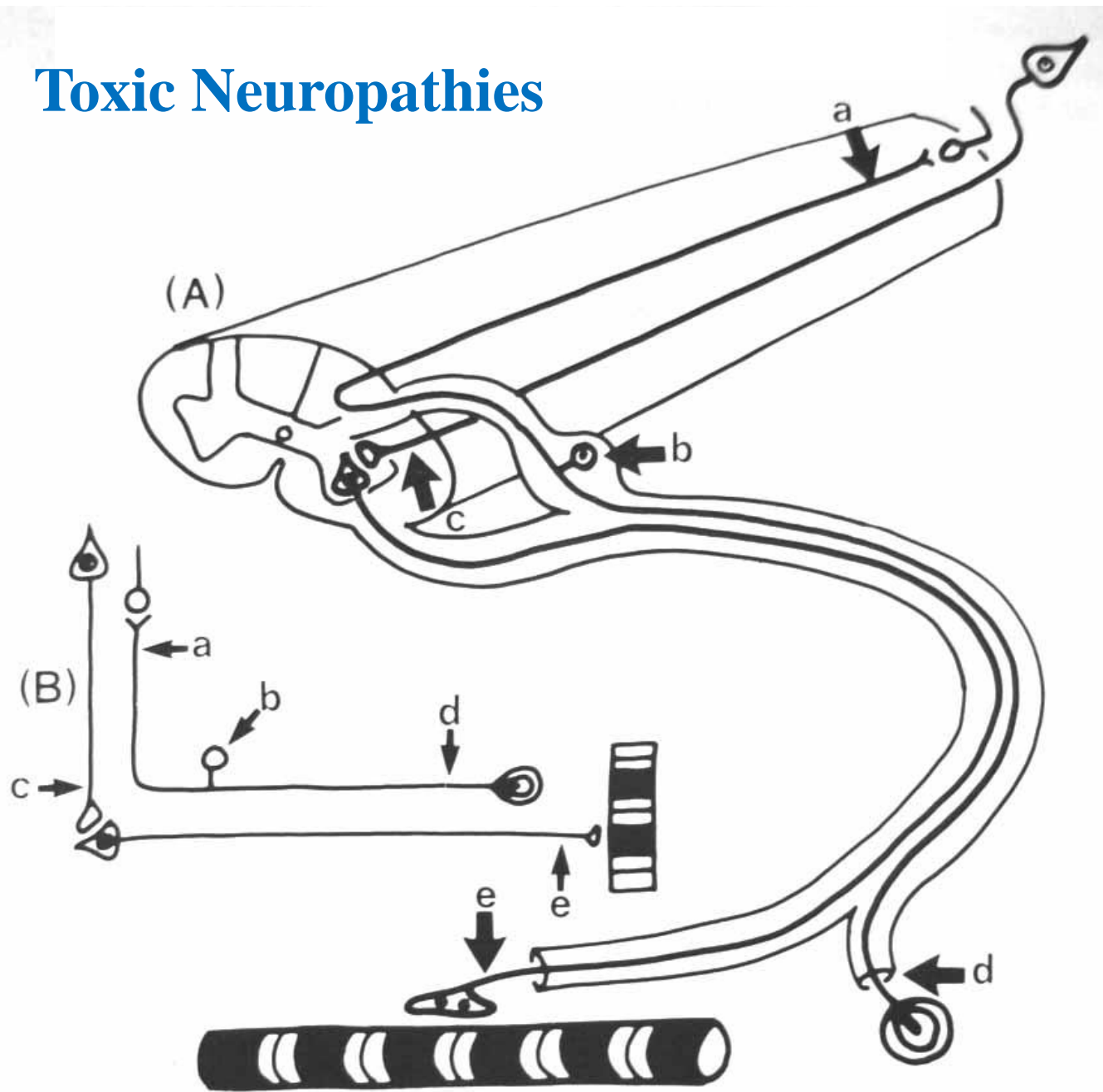


*F. Netter M.D.*  
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# Clinical Evaluation of Compression Neuropathy



# Toxic Neuropathies



# Nerve Biopsy

- **When to biopsy a nerve?**
  - **Neuropathy is severe, actively worsening**
  - **Essential for diagnosis**
- **Complications**
  - **10-15%**
  - **Localised Sensory loss**
  - **Wound infection, dehiscence, neuroma formation**
  - **Unpleasant dysaesthesiae, neuropathic pain**



# Peripheral Neuropathies in Which a Nerve Biopsy May Be Useful

- Acquired

- Vasculitis\*
- Sarcoidosis\*
- Amyloidosis\*
- CIDP
- IgM Paraproteinaemic N
- Leprosy
- Tumour infiltration\*

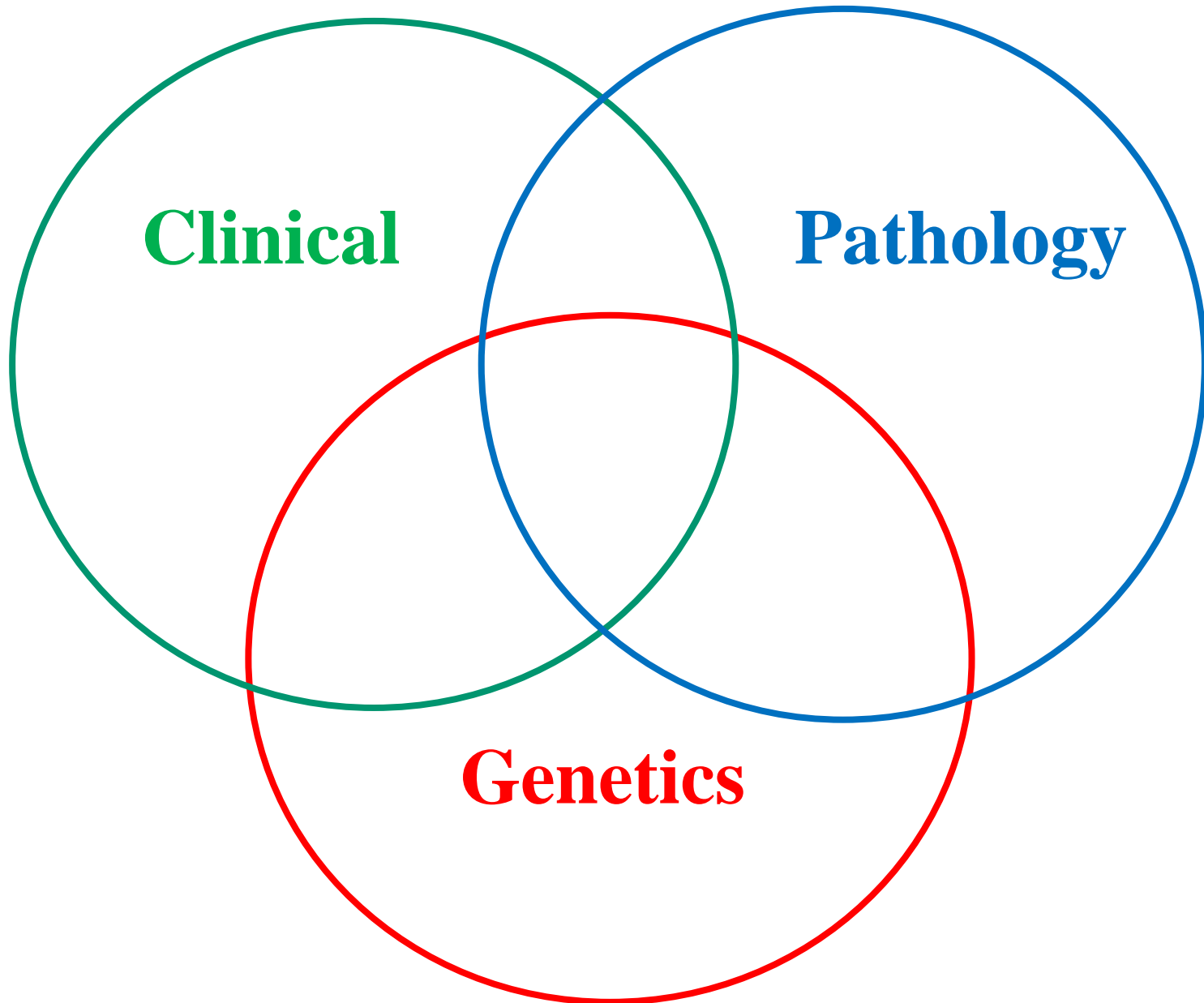
- Hereditary

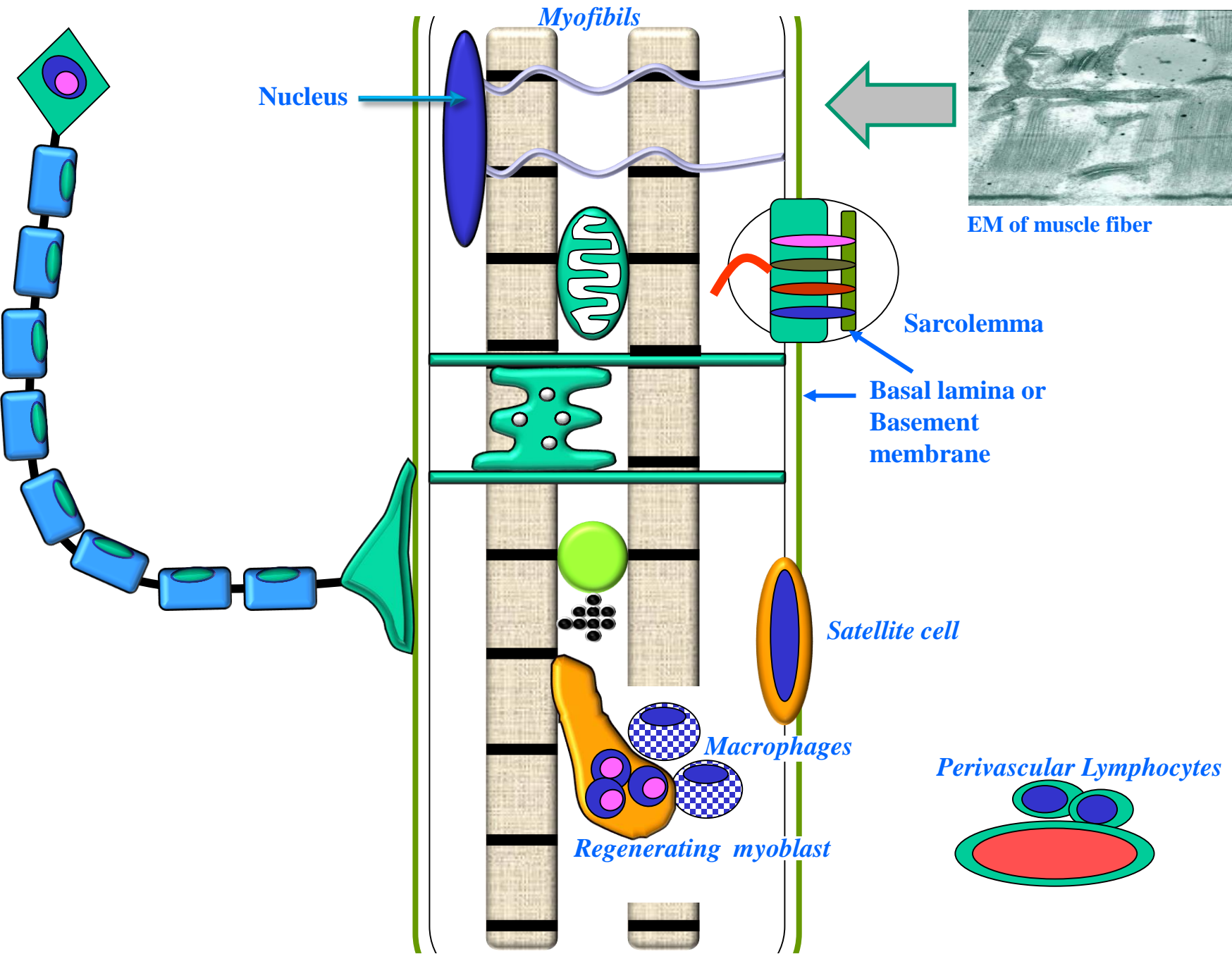
- CMT types 1A,1B &3
- HNPP (tomaculous neuropathy)
- Amyloidosis
- Giant Axonal Neuropathy
- Metachromatic Leukodystrophy
- Polyglucosan body N\*
- Refsum's disease

\*Nerve Biopsy often essential for Dx

# Muscle disease

# Diagnosis of Muscle Disease





# Clinical Classification of Myopathies: Primary diseases of Muscle

- Hereditary:

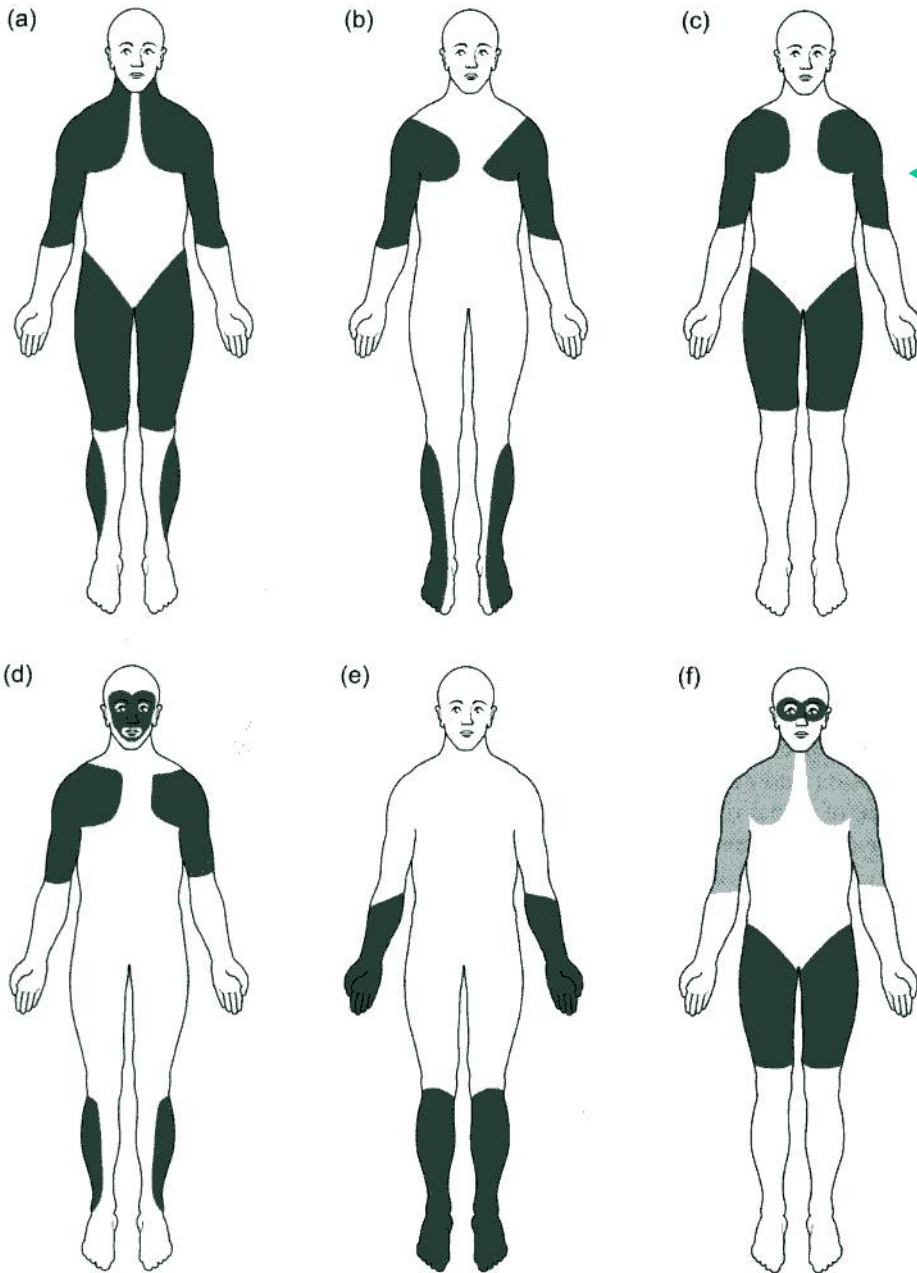
- Muscular dystrophies
- Myotonias
- Channelopathies
- Congenital myopathies
- Metabolic myopathies
- Mitochondrial myopathies

- Acquired:

- Inflammatory myopathies
- Endocrine myopathies
- Myopathies associated with other systemic illness
- Drug-induced myopathies
- Toxic myopathies

# Myopathy

- Features Supporting Diagnosis:
  - **Distribution**                      **proximal**
  - **Muscle Bulk**                      **preserved or enlarged**
  - **Reflexes**                              **Parallel muscle strength**



Most myopathies have a proximal distribution of weakness and pain.

**Distribution of weakness can aid diagnosis in muscular dystrophies:**

- a] Duchenne and Becker;
- b] Emery Dreifuss;
- c] limb-girdle;
- d] fascioscapulohumeral;
- e] distal;
- f] oculopharyngeal

# Myopathy

- Features against diagnosis:
  - Distal weakness
  - Fasciculations
  - Tremor
  - Sensory signs (or symptoms)
  - Pathological fatigue
  - Early absence of reflexes



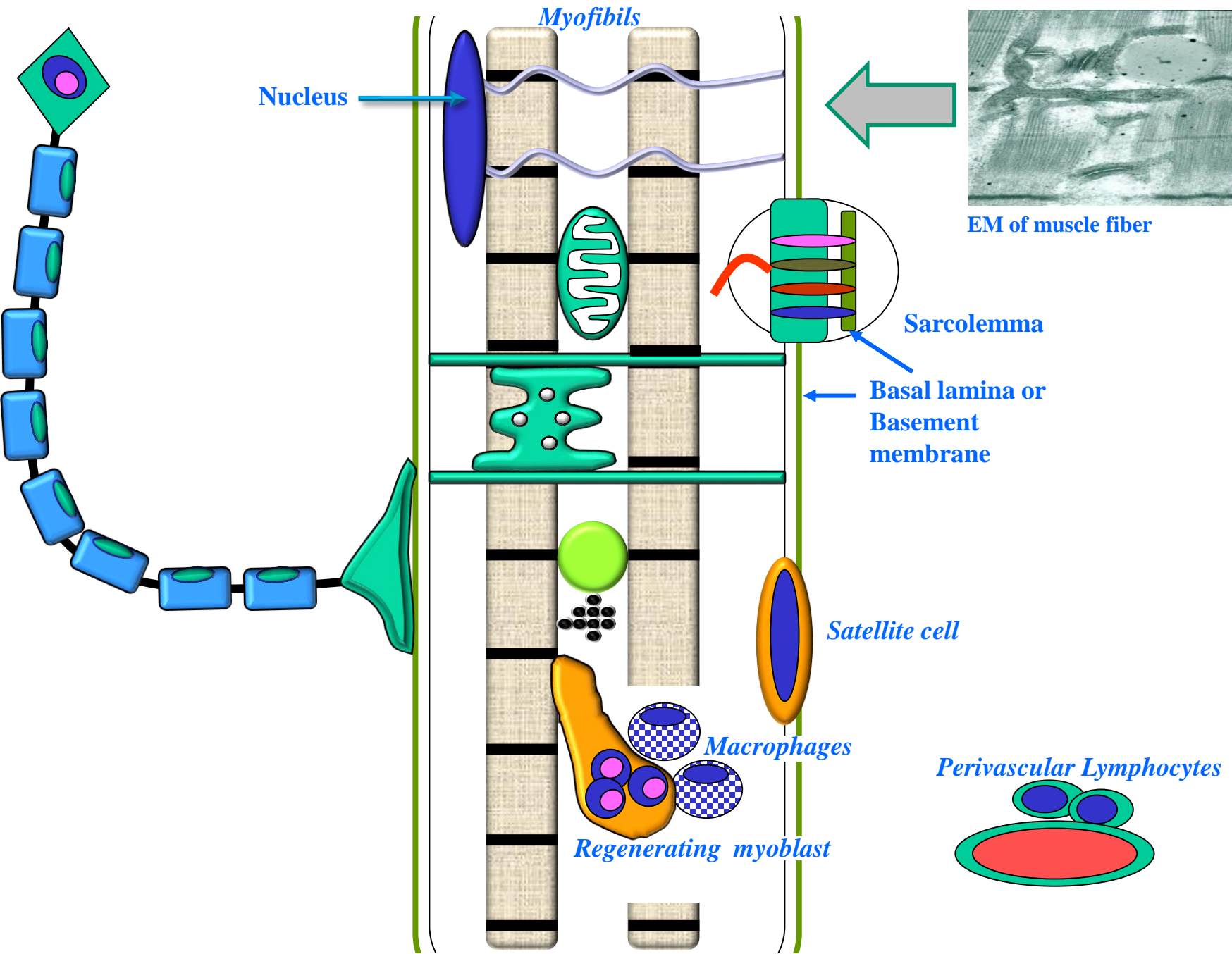
# Muscle biopsy

- Which Muscle?
  - Moderately involved, but avoid muscle with severe weakness
  - Best Specific Muscles: Deltoid, Biceps, Quadriceps
  - Avoid: Muscle sampled by EMG or sites of recent trauma

**Muscle Disease: Pathology &  
Genetics  
2<sup>nd</sup> Edition**

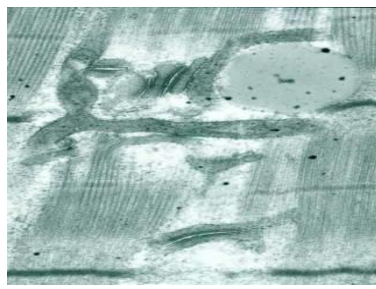
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*Myofibrils*

Nucleus



EM of muscle fiber

Sarcolemma

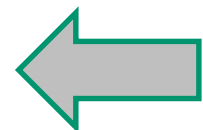
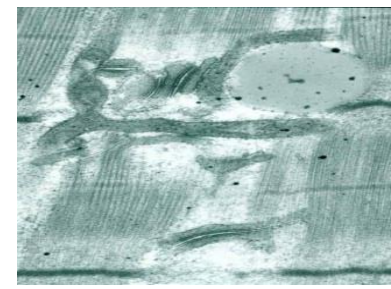
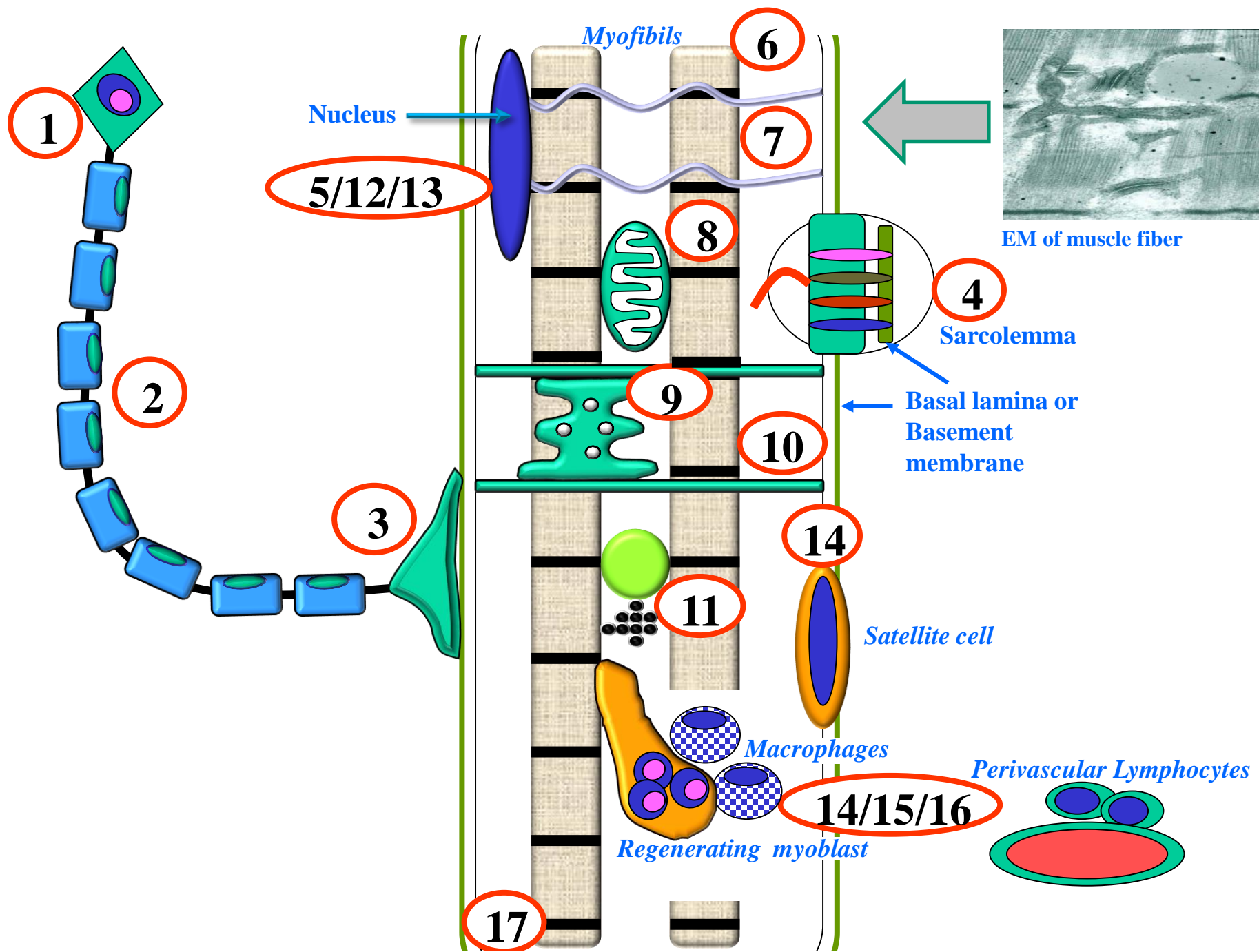
Basal lamina or  
Basement  
membrane

*Satellite cell*

*Macrophages*

*Perivascular Lymphocytes*

*Regenerating myoblast*



**14/15/16**

