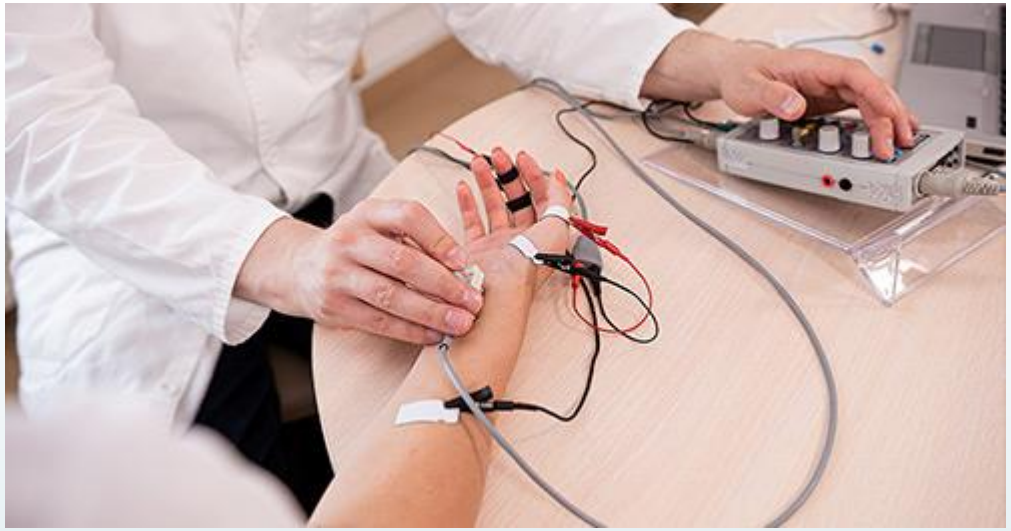

Approach to Nerve Conduction Studies and Electromyography

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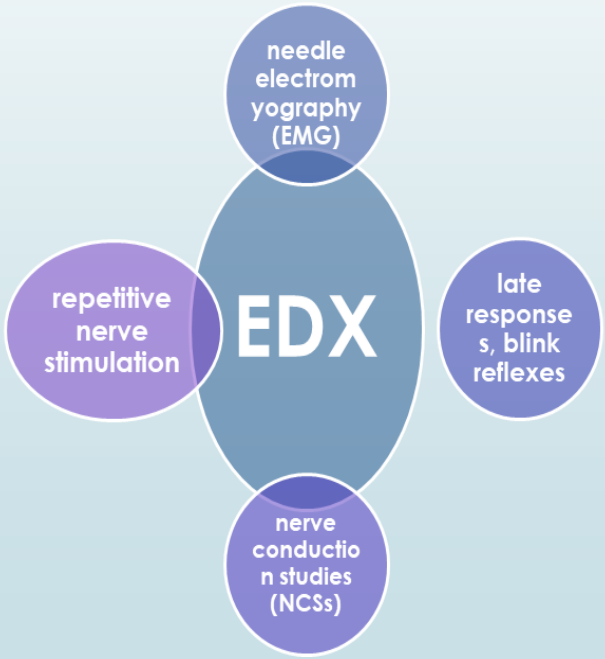
Introduction

- Electrodiagnostic (EDX) studies play a key role in the evaluation of patients with neuromuscular disorders. Among these studies are included:

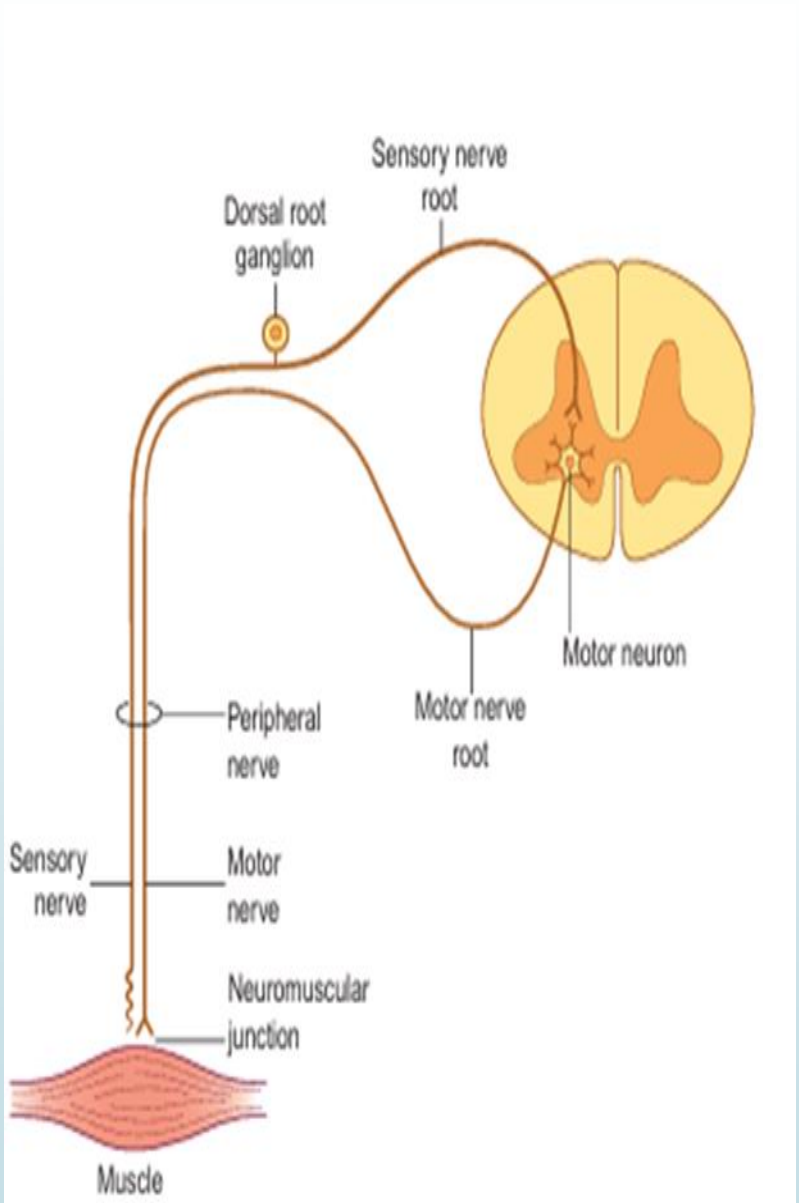


- **NCSs** and needle **EMG** form the core of the EDX study.
- They are performed first, and usually yield the greatest diagnostic information.
- NCSs and needle EMG are complementary and are always performed together and **during the same setting.**
- Performed and interpreted correctly, EDX studies yield critical information about the underlying neuromuscular disorder and allow the use of other laboratory tests appropriately and efficiently.
- Likewise, the information gained from EDX studies often leads to specific medical or surgical therapy.
- For example, a patient with a peripheral neuropathy clinically, who is subsequently found to have an acquired demyelinating neuropathy with conduction blocks on EDX studies, most often has a potentially treatable condition

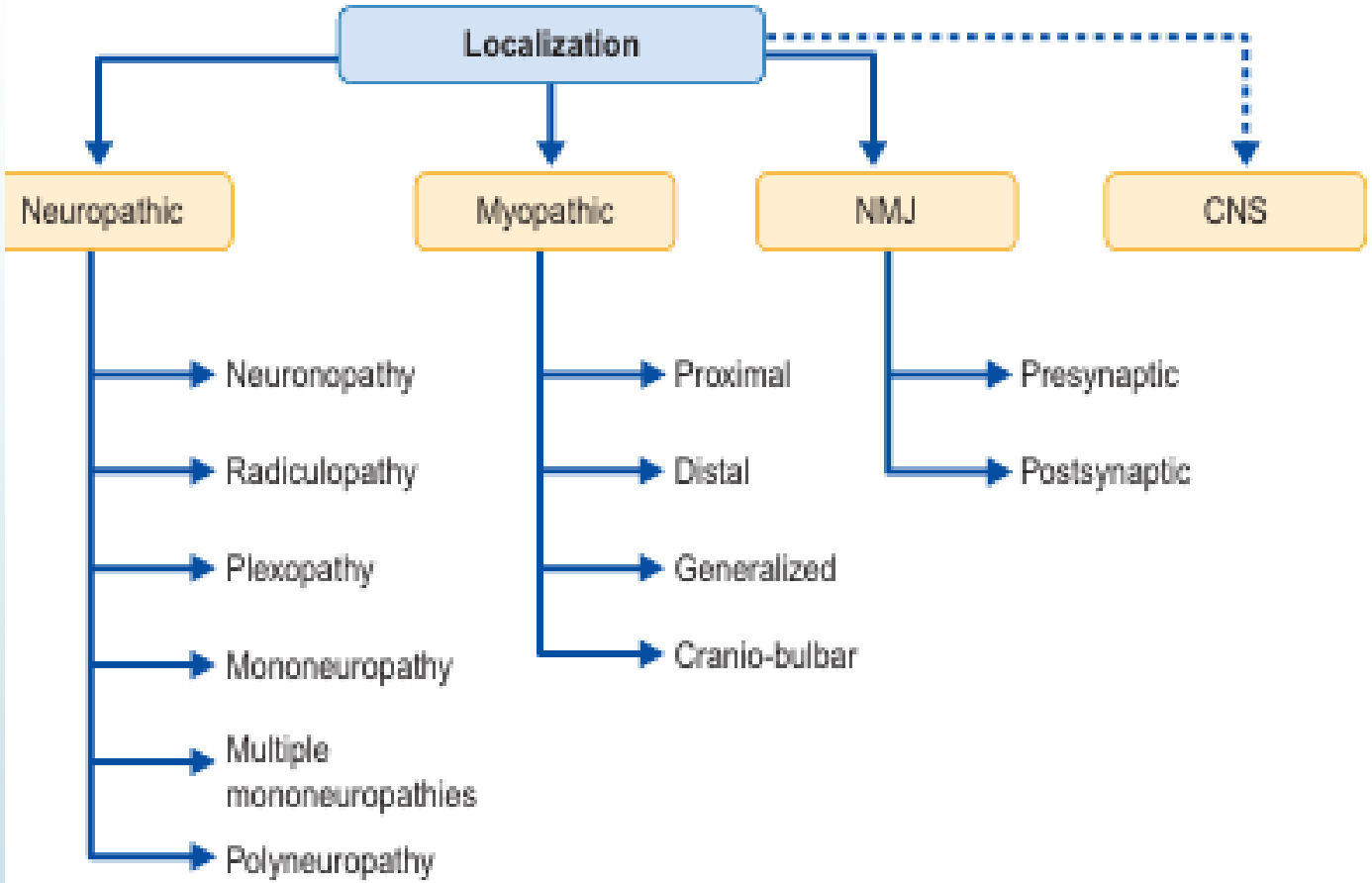
- NCSs and EMG are most often used to diagnose disorders of the peripheral nervous system (Figure 1–1, Box 1–1).
- These include disorders affecting the primary motor neurons (anterior horn cells), primary sensory neurons (dorsal root ganglia), nerve roots, brachial and lumbosacral plexuses, peripheral nerves, neuromuscular junctions, and muscles.
- In addition, these studies may provide useful diagnostic information when the disorder arises in the central nervous system (e.g., tremor or upper motor neuron weak ness). Occasionally, information from the EDX study is so specific that it suggests a precise etiology.

Box 1–1. Disorders of the Peripheral Nervous System

- Motor neuronopathy
  - Amyotrophic lateral sclerosis
  - Spinal muscular atrophy
  - Infectious (poliomyelitis, West Nile virus)
  - Monomelic amyotrophy
- Sensory neuronopathy
  - Paraneoplastic
  - Autoimmune
  - Toxic
  - Infectious
- Radiculopathy
  - Disk herniation
  - Spondylosis
  - Neoplastic
  - Infarction
  - Infectious
  - Inflammatory
- Plexopathy
  - Radiation induced
  - Neoplastic
  - Entrapment
  - Diabetic
  - Hemorrhagic
  - Inflammatory
- Neuropathy
  - Entrapment
  - Polyneuropathy
    - Demyelinating
    - Axonal
  - Mononeuritis multiplex
- Neuromuscular junction disorders
  - Myasthenia gravis
  - Lambert–Eaton myasthenic syndrome
  - Botulism
  - Toxic
  - Congenital
- Myopathy
  - Inherited
    - Muscular dystrophy
    - Congenital
    - Metabolic
  - Acquired
    - Inflammatory
    - Toxic
    - Endocrine
    - Infectious

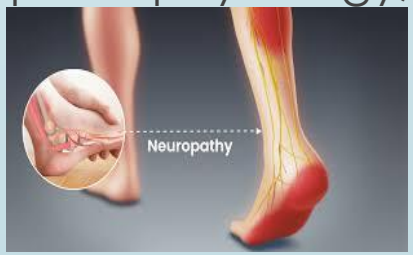


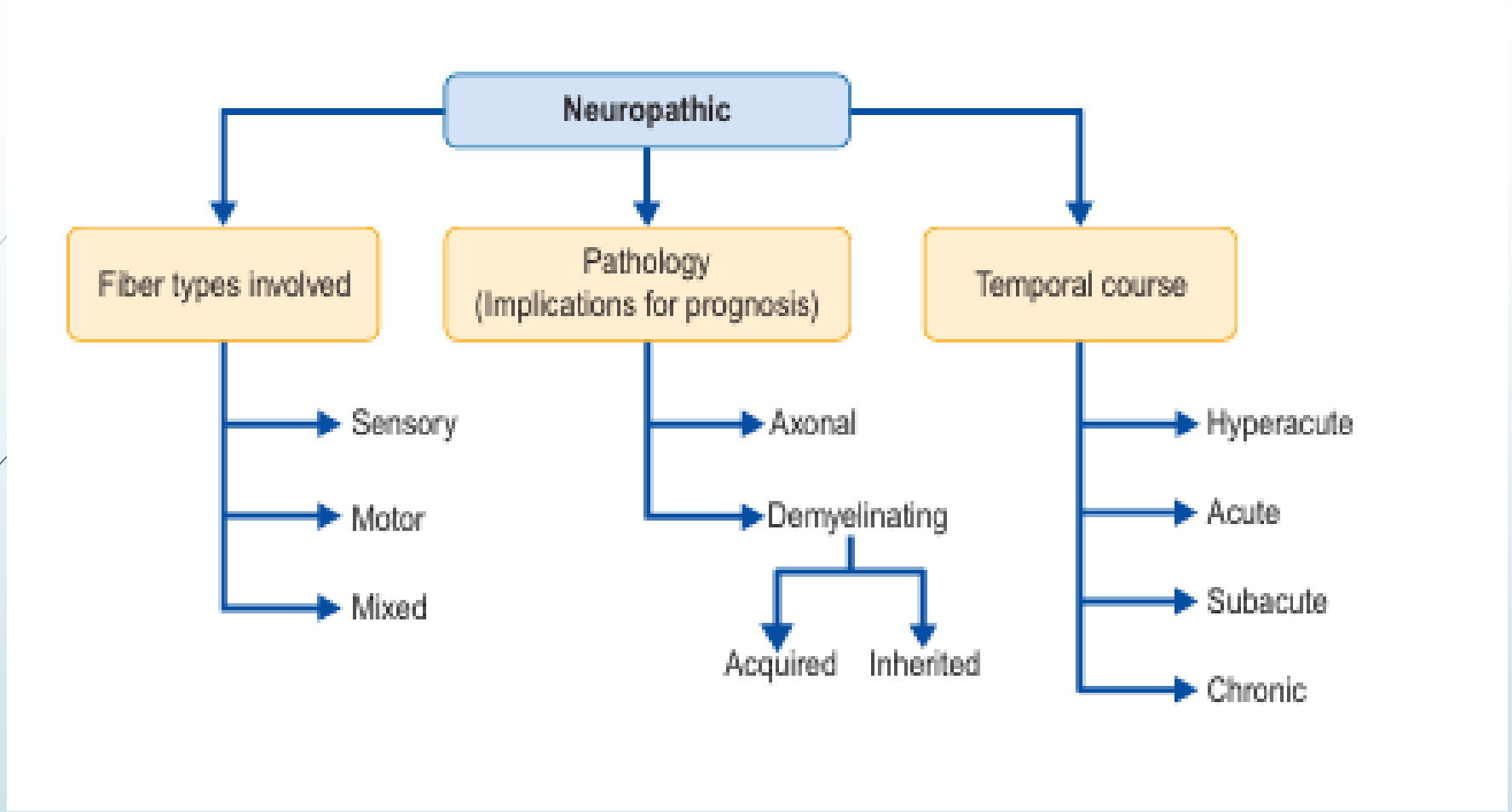
LOCALIZATION OF THE DISORDER IS THE MAJOR AIM OF THE ELECTRODIAGNOSTIC STUDY


Localization
Neuropathic
Myopathic
NMJ
CNS
Neuronopathy
Radiculopathy
Plexopathy
Mononeuropathy
Multiple mononeuropathies
Polyneuropathy
Proximal
Distal
Generalized
Cranio-bulbar
Presynaptic
Postsynaptic

Neuropathic Localization

- ***Neuropathic*** is probably the most common localization made on EDX studies.
- ***Neuropathic*** literally means a disorder of the peripheral nerves. However, in common usage, it includes the primary sensory and motor neurons as well.
- EDX studies are particularly helpful in neuropathic conditions. First, in conjunction with the history and examination, they can usually further localize the disorder to the neurons, roots, plexus, or peripheral nerve.
- In the case of peripheral nerve, further localization is usually possible to a single nerve (mononeuropathy), multiple individual nerves (mononeuropathy multiplex) or all nerves (polyneuropathy). In the case of a single nerve, the exact segment of nerve responsible for the problem may be localized in some cases.
- In the case of neuropathic lesions, EDX studies often yield further key information, including the fiber types involved, the underlying pathophysiology, and the temporal course of the disorder




Neuropathic
Fiber types involved
Pathology
(Implications for prognosis)
Temporal course
Sensory
Motor
Mixed
Axonal
Demyelinating
Acquired
Inherited
Hyperacute
Acute
Subacute
Chronic

Information About the Fiber Types Involved and the Underlying Nerve Pathophysiology can be Gained, which then Further Narrows the Differential Diagnosis

- First, EDX studies are more sensitive than the clinical examination in determining which fiber types are involved: motor, sensory, or a combination of the two.
- Sensorimotor polyneuropathies are common and suggest a fairly large differential diagnosis.
- On the other hand, pre dominantly motor or predominantly sensory neuropathies are rare and suggest a much more limited set of disorders.
- For instance, a patient with numbness in the hands and feet and diminished reflexes may be diagnosed with a peripheral neuropathy.
- However, if EDX studies demonstrate abnormal sensory nerve conductions with completely normal motor nerve conductions and needle EMG, then the differential diagnosis changes from a peripheral neuropathy to a pure sensory neuropathy or neuronopathy, which has a much more limited differential diagnosis.

- **Second,** EDX studies often can define whether the underlying pathophysiology is demyelination or axonal loss.
- Although most demyelinating neuropathies have some secondary axonal loss and many axonal loss neuropathies have some secondary demyelination, EDX studies usually can differentiate between a primary demyelinating and a primary axonal neuropathy.
- Because EDX studies usually can make this differentiation quickly and non-invasively, nerve biopsy is essentially never required to make this determination. Furthermore, the differentiation between primary axonal and primary demyelinating pathology is of considerable diagnostic and prognostic importance, especially in the case of polyneuropathies.
- The vast majority of polyneuropathies are associated with primary axonal degeneration, which has an extensive differential diagnosis. In contrast, the number of true electrophysiologic primary demyelinating neuropathies is extremely small.

Assessing the Degree of Axonal Loss versus Demyelination has Implications for Severity and Prognosis

- A nerve that has sustained a demyelinating injury often can remyelinate in a very short time, usually weeks. However, if there has been substantial axonal loss, whether primary or secondary, the prognosis is much more guarded.
- The rate of axonal regrowth is limited by the rate of slow axonal transport, approximately 1 mm per day.
- Clinically, axonal loss lesions can rarely be differentiated from demyelinating ones, especially in the acute setting. For example, in a patient who awakens with a complete wrist and finger drop, the etiology usually is compression of the radial nerve against the spiral groove of the humerus. However, the paralysis could result from either conduction block (i.e., demyelination) or axonal loss, depending on the severity and duration of the compression. Clinically, both conditions appear the same. Nevertheless, if the injury is due to axonal loss, it has a much worse prognosis and a longer rehabilitation time to recovery than a similarly placed lesion that is predominantly demyelinating in nature. EDX studies can readily differentiate axonal from demyelinating lesions.

Assessment of the Temporal Course can Often be Made

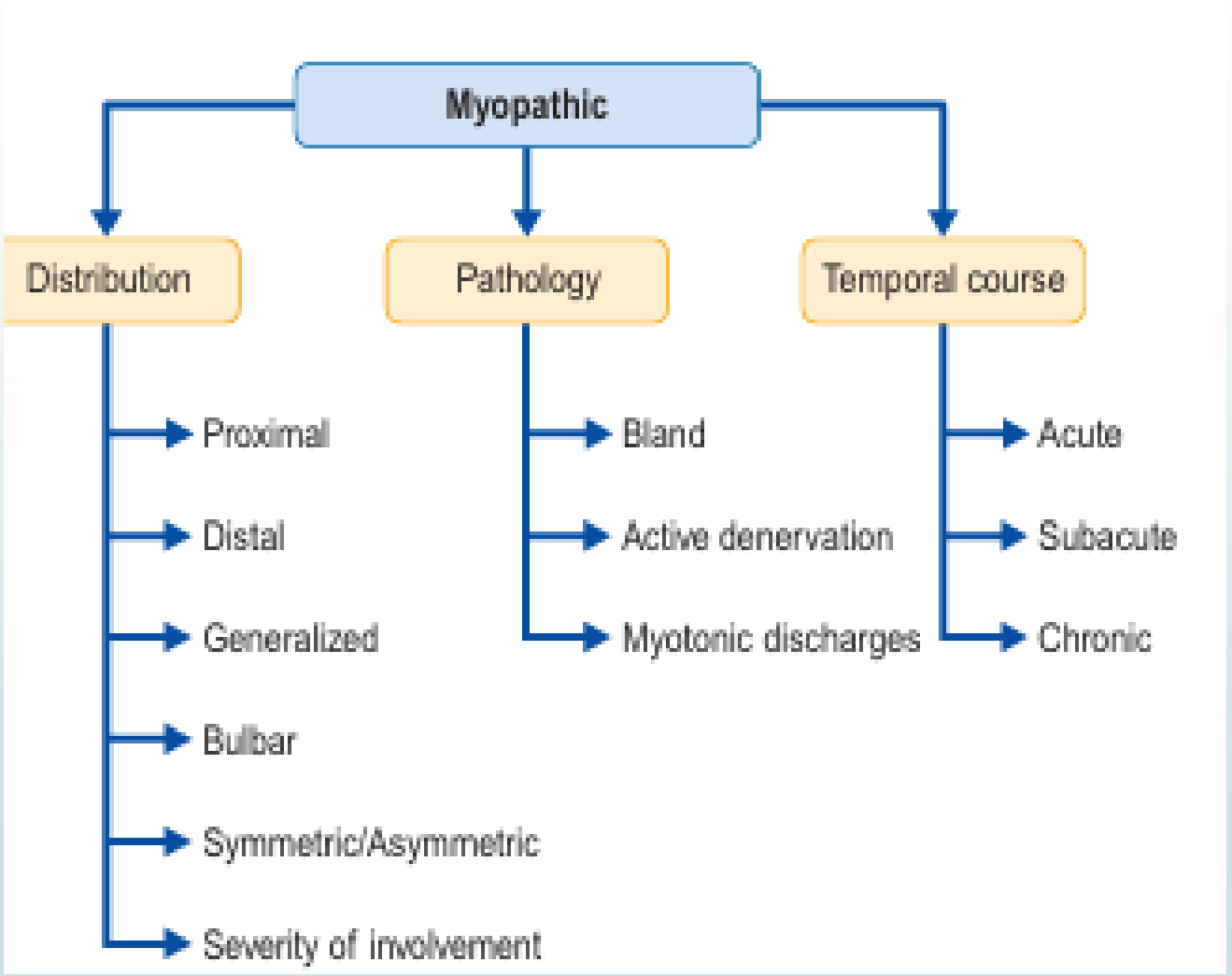
- For neuropathic conditions, there is an orderly, temporal progression of abnormalities that occurs in NCSs and needle EMG.
- A combination of findings often allows differentiation among hyperacute (less than one week), acute (up to a few weeks), subacute (weeks to a few months), and chronic (more than a few months) lesions.
- The time course suggested by the EDX findings may alter the impression and differential diagnosis. For example, it is not uncommon for a patient to report an acute time course to his or her symptoms, whereas the EDX studies clearly indicate that the process has been present for a longer period of time than the patient has been aware of.

- Conversely, the temporal course described by the patient may impact the interpretation of the EDX findings.
- For instance, the finding of a normal ulnar sensory nerve action potential recording the little finger, in a patient with numbness of the little finger, has very different implications depending on the time course of the symptoms.
- If the symptoms are truly less than one week in duration, the normal ulnar sensory response could indicate an ulnar neuropathy (with incomplete Wallerian degeneration), a proximal demyelinating lesion, or a lesion at the level of the nerve root or above.
- On the other hand, if the symptoms have been present for several weeks or longer, the same finding would indicate either a proximal demyelinating lesion or a lesion at the level of the nerve root or above.

Myopathic Localization

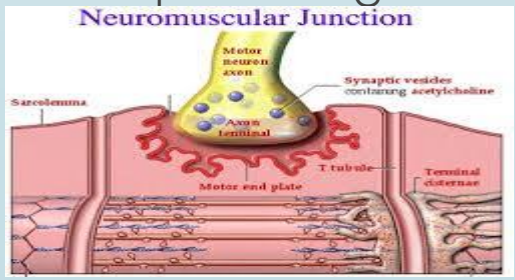
- In the case of myopathic (i.e., muscle) disease, EDX studies can also add key information to further define the condition (Figure 1–4).
- First, the distribution of the abnormalities may suggest a particular diagnosis: are they proximal, distal or generalized? Most myopathies preferentially affect proximal muscles.
- Few myopathies, such as myotonic dystrophy type I, affect distal muscles.
- Some very severe myopathies (e.g., critical illness myopathy) can be generalized.
- In rare myopathies, there is prominent bulbar weakness; accordingly, EDX abnormalities may be most prominent in the bulbar muscles.
- Most myopathies are fairly symmetric; the finding of asymmetry either clinically and/or on EDX studies can be very helpful in narrowing the differential diagnosis. For example, inclusion body myositis may present asymmetrically, whereas polymyositis and dermatomyositis do not.

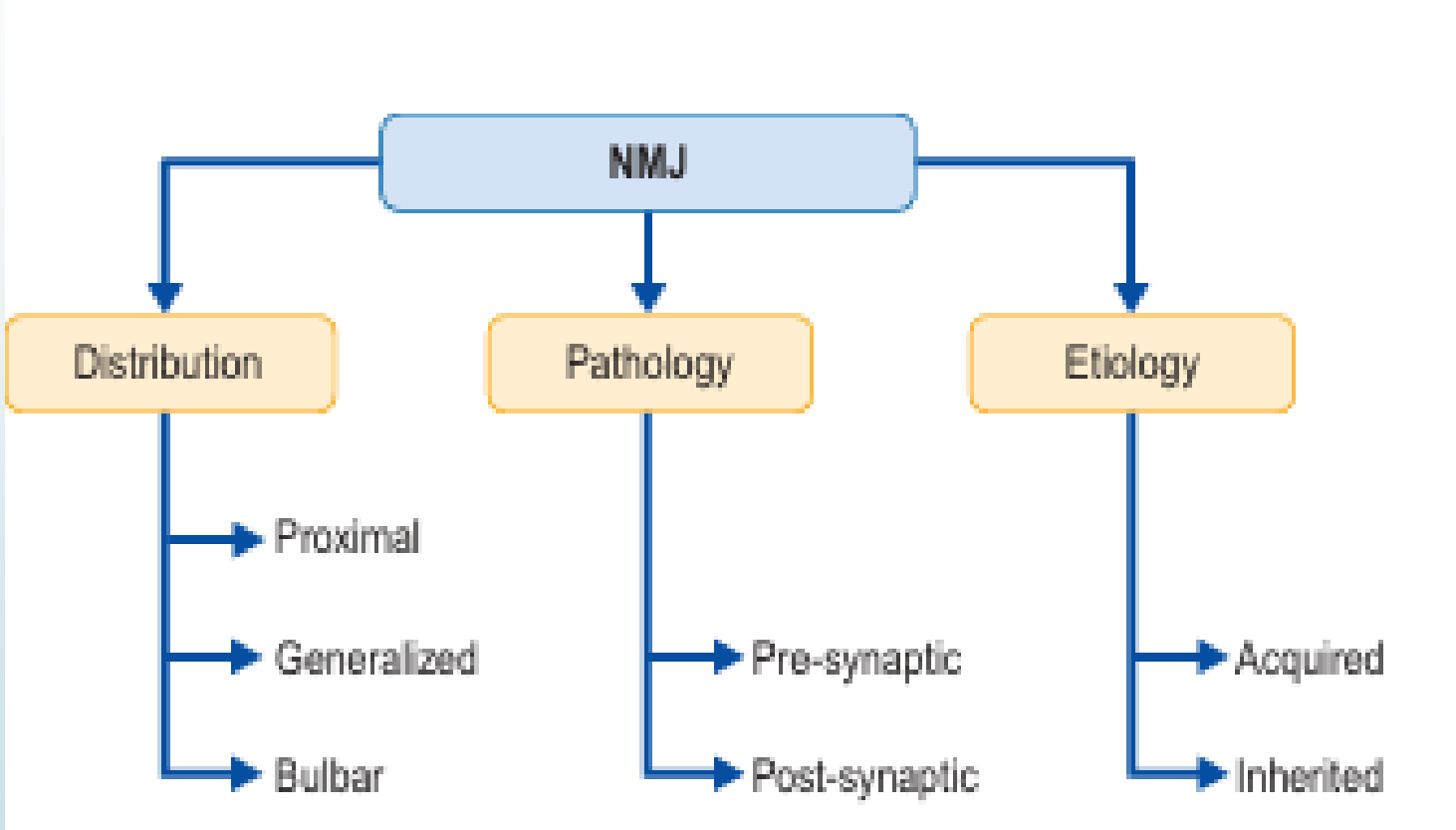
- Second, the presence of spontaneous activity on needle EMG is helpful in limiting the differential diagnosis and suggesting certain underlying pathologies.
- Most myopathies are bland with little or no spontaneous activity.
- However, myopathies which are inflammatory, necrotic and some which are toxic may be associated with active denervation.
- In addition, other myopathies may have prominent myotonic discharges at rest. The presence of myotonic discharges in a myopathy markedly narrows the differential diagnosis to only a few possible disorders.
- Lastly is the issue of the temporal course. Although this determination is more challenging than with neuropathic lesions, in some myopathies, a determination can be made if the myopathy is acute, subacute, or chronic, a finding which again narrows the differential diagnosis


Myopathic
Distribution
Pathology
Temporal course
Proximal
Distal
Generalized
Bulbar
Symmetric/Asymmetric
Severity of involvement
Bland
Active denervation
Myotonic discharges
Acute
Subacute
Chronic

Neuromuscular Junction Localization

- Disorders of the neuromuscular junction (NMJ) are distinctly uncommon.
- However, when they occur, EDX studies not only help in identifying them, but can add other key pieces of information (Figure 1–5).
- First is the distribution of the abnormalities on EDX testing: are they proximal, bulbar or generalized? For instance, myasthenia gravis preferentially affects oculobulbar muscles and then proximal muscles on EDX studies, whereas myasthenic syndrome is a generalized disorder on EDX studies, although clinically it has a predilection for proximal muscles.
- Broadly speaking, the underlying pathology can be divided into pre-synaptic and post-synaptic disorders. EDX studies are usually very good at making this determination. Myasthenia gravis is the prototypic post-synaptic disorder, whereas myasthenic syndrome and botulism target the pre synaptic junction.
- Lastly is the issue of the etiology of the NMJ disorder, whether it is acquired or inherited. Almost all NMJ disorders are acquired. However, there are rare inherited NMJ disorders. In some of these, there may be unique findings on EDX testing that suggest one of these rare disorders.




NMJ
Distribution
Pathology
Etiology
Proximal
Generalized
Bulbar
Pre-synaptic
Post-synaptic
Acquired
Inherited

PATIENT ENCOUNTER

before starting every study, the EDX physician must know some basic facts:

- What are the patient's symptoms?
- How long have they being going on?
- Is there any important past medical history (e.g., diabetes, history of chemotherapy, etc.)?
- Is there muscle atrophy?
- What is the muscle tone (normal, decreased or increased)?
- Is there weakness and, if so, where is it and how severe is it?
- What do the reflexes show (normal, decreased or increased)?
- Is there any loss of sensation and, if so, what is the distribution; what modalities are disturbed (e.g., temperature, pain, vibration, etc.)?

Box 1–2. Patient Encounter

1. Take a brief history and perform a directed physical examination
2. Formulate a differential diagnosis
3. Formulate a study based on the differential diagnosis
4. Explain the test to the patient
5. Perform the nerve conduction studies and modify which nerve conduction studies to add based on the findings as the test proceeds
6. Perform the needle electromyography study and modify which additional muscles to sample, based on the findings as the test proceeds

CARDINAL RULES OF NERVE CONDUCTION STUDIES AND ELECTROMYOGRAPHY

1. NCSs and EMG are an extension of the clinical examination.

- NCSs and EMG cannot be performed without a good clinical examination. Every examination must be individualized based on the patient's symptoms and signs and the resulting differential diagnosis. If marked abnormalities are found on electrophysiologic testing in the same distribution where the clinical examination is normal, either the clinical examination or the electrophysiologic testing must be called into question.
- One usually finds that the better the clinical examination, the better the differential diagnosis, and thus the more clearly directed the EDX studies will be.

- 2. When in doubt, always think about technical factors. EDX studies rely upon collecting and amplifying very small bioelectric signals in the millivolt and microvolt range. Accomplishing this is technically demanding; a large number of physiologic and non-physiologic factors can significantly interfere with the accuracy of the data.
- Technical problems can easily lead to absent or abnormal findings. Failure to recognize technical factors that influence the EDX study can result in **type I errors** (i.e., diagnosing an abnormality when none is present), and **type II errors** (i.e., failing to recognize an abnormality when one is present).

- 3. **When in doubt, reexamine the patient**.
- This is essentially an extension of cardinal rule number 1. In the example given with rule number 2, if the sural sensory response is absent after all possible technical factors have been corrected, the clinician should reexamine the patient. If the patient has clear loss of vibration at the ankles, there is less concern about an absent sural sensory response. If the patient's sensory examination is normal on reexamination, the absent sensory response does not f it the clinical findings, and technical factors should be investigated further

- 4. **EDX findings should be reported in the context of the clinical symptoms and the referring diagnosis.** In every study, electrophysiologic abnormalities must be correlated with the clinical deficit. Because electrophysiologic studies are quite sensitive, it is not uncommon for the electromyographer to discover mild, subclinical deficits of which the patient may not be aware. For example, a diabetic patient referred to the EMG laboratory for polyneuropathy may show electrophysiologic evidence of a superimposed ulnar neuropathy but have no symptoms of such. Accordingly, the electromyographer should always report any electrophysiologic abnormality in the context of its clinical relevance so that it can be properly interpreted.

- **When in doubt, do not overcall a diagnosis.**
- Because electrophysiologic tests are very sensitive, mild, subclinical, and sometimes clinically insignificant findings often appear on EDX testing.
- This occurs partly because of the wide range of normal values, which vary with the nerve and muscle being tested.
- In addition, there are a variety of physiologic and non-physiologic factors that may alter the results of both NCSs and EMG, despite attempts to control for them. These factors, often when combined, may create minor abnormalities.
- Such minor abnormalities should not be deemed relevant unless they correlate with other electrophysiologic findings and, most importantly, with the clinical history and examination. It is a mistake to overcall an electrophysiologic diagnosis based on minor abnormalities or on findings that do not fit together well. Sometimes, the clinical or electrophysiologic diagnosis is not clear-cut and a definite diagnosis cannot be reached

- ٦. **Always think about the clinical–electrophysiologic correlation**. \
- This rule combines all of the earlier rules. One usually can be certain of a diagnosis when the clinical findings, NCSs, and EMG abnormalities all correlate well. Consider again the example of the patient with weakness of the hand and tingling and numbness of the fourth and fifth fingers. If NCSs demonstrate abnormal ulnar motor and sensory potentials associated with slowing across the elbow, and the needle EMG shows denervation and reduced numbers of motor unit potentials in all ulnar innervated muscles and a normal EMG of all non ulnar-innervated muscles, there is a high degree of certainty that the patient truly has an ulnar neuropathy at the elbow, and the electrophysiologic abnormalities are indeed relevant.

Thank you!