ELECTRODIAGNOSTIC (EDX) STUDIES IN AN INTENSIVE CARE UNIT (ICU): NEUROLOGICAL PROBLEMS, RECOMMENDED EDX PROTOCOL AND COMMON EDX PATTERNS AT A GLANCE.

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Abstract In past several years requests for intensive care unit EDX studies for patients with profound illness and overlapping medical problems have increased. Issues like shifting these cases from ICU to EDX laboratory becomes major hurdle thereby generating a need for portable and bedside EDX study. EDX studies are most often requested in ICU for patients presenting with rapidly progressive weakness with or without motor or sensory symptoms, often leading to respiratory compromise and intubation. Secondly, when a patient is recovering from non neurologic problem where he or she required intubation and sedation, drugs are weaned and we observe profound weakness of limbs with hypotonia and areflexia. Thirdly, despite apparently intact cardiac and pulmonary function, patient fails to wean off the ventilator thus creating a dilemma for possible neuromuscular disorder that is preventing extubation. As an Intensivist we must remain updated about most common neurological issues that require specific neurophysiological studies and its electrodiagnostic patterns in intensive care unit. Current review highlights some important neurological issues that present with weakness and utility of EDX studies in delineating the lesion.

Key Words
Electrodiagnosis, Intensive care unit, Nerve conduction studies (NCS), Electromyography (EMG)

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Intensive care physicians often need input from neurophysicians to deal with the management of primary disease of central and peripheral nervous system which may cause encephalopathy, ventilator, autonomic and bulbar insufficiency or profound neuromuscular weakness. [(1)] Neurologic causes of profound weakness in an ICU patient include disorders of central nervous system (CNS) and the peripheral nervous system (PNS). Weakness may occur due to primary neurological conditions or occurs while patient is admitted for some other medical conditions. These patients often present as difficulty in weaning from ventilation, reduced movements in an obtunded patient or generalized or local weakness in an awake and alert patient. [(2, 3)]

Central nervous system disorders

Most common CNS diagnosis that leads to weakness in ICU is encephalopathy. It has multifactorial origin including metabolic and electrolyte imbalance, sepsis and medications. Septic encephalopathy is the most common form of encephalopathy encountered in intensive care medicine. [(4)] Stroke, anoxia, seizures, subarachnoid hemorrhage and infectious meningitis are other neurological problems presenting with weakness in ICU. Spinal cord disorders like infarction demyelination or trauma at high cervical cord level can also present as generalized weakness. [(5)]

Peripheral nervous system disorders

Among the list of PNS disorders that presents with profound weakness, lesion may be anywhere in lower motor neuron from anterior horn cell, its axon, neuromuscular junction or muscle. Acute motor neuron disease is very uncommon and occurs only with paralytic poliomyelitis. Chronic motor neuron disease such as amyotrophic lateral sclerosis (ALS) occasionally may land in ICU with pneumonitis and respiratory failure if not appreciated previously due to its bulbar presentation. It is only when patient is recovering from pneumonia and trial for extubation fails, EDX studies gives ALS picture that was not recognized earlier. [(6, 7)]

A well known acute neuropathy with marked weakness and respiratory failure is Guillain-Barre` syndrome. It is an acquired inflammatory motor and sensory polyradiculoneuropathy with a probable autoimmune etiology. It is very rare occasion to see an acute neuropathy other than GBS in an ICU. Notable exceptions include porphyria and some toxic (arsenic) neuropathies that can mimic presentation of GBS. [8, 9]

Critical illness polyneuropathy (CIP) is most common severe neuropathy seen in ICU patients admitted for medical illness with multiple organ failure and sepsis. [10] It is an axonal sensorimotor polyneuropathy, thought to be due to a complication of systemic inflammatory response (septic) syndrome (SIRS). This syndrome occurs in response to severe infection or trauma due to invasive procedures in ICU patients hospitalized for more than a week. It is noticed only when patient begins to improve from primary illness but is found to have profound weakness with sensory loss or fails to wean from the ventilator. [11] Phrenic mononeuropathy is another reason for respiratory compromise. It may be unilateral or bilateral, idiopathic or traumatic (thoracic surgeries). [12]

Among the list of neuromuscular junction (NMJ) disorders in ICU are those presenting with rapidly progressive weakness (acute) and those with subacute presentation over months. Botulism, organophosphate poisoning, spider venom and 'nerve gas' are few acute presentations. [13] Occasionally, a myasthenia gravis (MG) may present to ICU with primary respiratory failure if diaphragm and or other respiratory muscles are selectively involved. Patients with Lambert-Eton myasthenic syndrome (LEMS) are distinctly uncommon in ICU and may mimic clinically as myopathy. LEMS presents as failure to wean after an elective surgery wherein patients were administered calcium channel blockers or neuromuscular blocking agents (NMBA) that might have unmasked the disease. [14, 15]

Most common muscle disorder seen in ICU is critical illness myopathy (CIM) also known as intensive care myopathy. It develops when high dose intra venous steroids are combined with NMBA but can also occur following major organ transplantation, especially liver. One of the most common situations where CIM occurs is in patients with status asthmaticus that are intubated (NMBA used) and administered with high dose methylprednisolone. Asthma improves but it is observed that patient is flaccid, areflexic with profound weakness. It may take prolonged period for extubation. Other myopathies that may occasionally present in ICU with respiratory arrest or generalized weakness are inflammatory myopathy, toxic myopathy, hypokalemic periodic paralysis and adult-onset acid maltase deficiency. [16, 17, 18]

Recommended EDX study protocol in the intensive care unit [5]

An electromyographer has to face many technical problems while performing an EDX study in ICU. Poor patient cooperation, non accessibility to some nerves and muscles due to position of patient and IV lines, excessive electrical noise etc. Irrespective of these problems an electromyographer must try his best to acquire accurate data and collect sufficient EDX evidences to reach the diagnosis. Intensivist must also get all the specific EDX studies done before finalizing the diagnosis. Recommended nerve conduction studies, repetitive nerve stimulation and electromyography protocol that should be well kept in mind according to differential diagnosis is as follows.

Routine NCS

1. At least one motor nerve conduction study with its corresponding F wave in upper and lower extremity.
2. At least one sensory nerve conduction study in upper and lower extremity.

Routine needle EMG

1. Lower extremity: at least one distal and one proximal muscle.

2. Upper extremity: at least one distal and one proximal muscle.

Special considerations: if adult-onset acid maltase deficiency is in the differential diagnosis, sampling the paraspinal muscle is essential.

Repetitive nerve stimulation (RNS)

1. Routine slow (3-Hz) repetitive nerve stimulation in at least one nerve
2. In any patient with absent borderline or low compound muscle action potential (CMAP) amplitudes, exercise for 10 seconds and repeat RNS study to look for abnormal incremental response (rise in CMAP amplitude). In case patient is not cooperative with voluntary exercise, stimulate motor nerve with 50-Hz to look for an abnormal increment.

Other useful studies in selected situations

1. Direct muscle stimulation to differentiate between CIM and CIP.
2. Phrenic motor study (bilateral) to assess integrity of phrenic nerve.

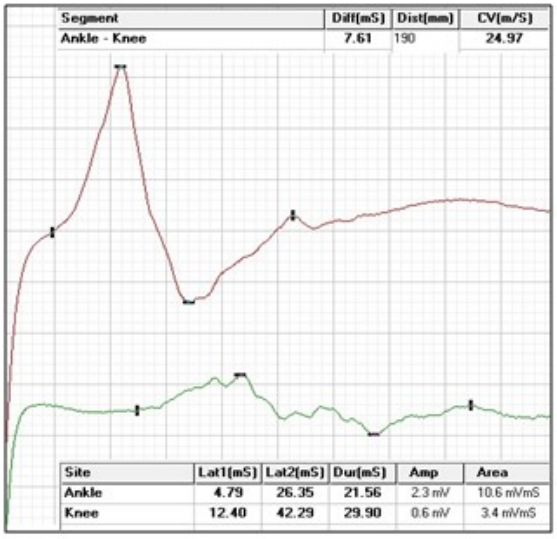
Important EDX patterns in the intensive care unit (ICU)

Very few EDX patterns are observed in an ICU depending upon the neurologic conditions resulting in respiratory or generalized weakness or both. Each pattern localizes the lesion and may guide us for additional studies if required. Various disorders and its EDX patterns are as follows.

1. Encephalopathy or other central nervous disorders: NCS studies (motor, sensory and RNS remains normal. F waves may be absent if patient is sedated or in coma. EMG shows poor activation. [(5)]
2. Motor neuron disease (ALS): Motor NCS shows reduced CMAPs (axonal loss) or normal CMAP amplitude. SNC study within normal limits. Normal or rarely decremental RNS response on slow (3Hz) stimulation. Spontaneous activity (SP activity) with fasciculations, positive sharp waves and fibrillations present. Decreased recruitment and activation of MUPs with signs of reinnervation (Long amplitude and duration, Polyphasic MUPs). [(19)]
3. Poliomyelitis: Reduced or normal CMAPs amplitude, normal SNC, RNS study and EMG shows reduced recruitment of MUPs in first week followed by active dennervation (SP activity present) and then reinnervation (Long polyphasic MUPs) [(20)]
4. AIDP or GBS: Depending on variant, NCS picture may vary. Mostly demyelinating features with slowed conductions (prolonged DML and reduced CV), conduction blocks and temporal dispersions at sites other than compressive mononeuropathies. Absent or prolonged latency F waves. Sensory spared in acute motor axonal neuropathy (AMAN) whereas initially normal but later with reduced SNAPs, slowed velocities in demyelinating, sensorimotor neuropathies. EMG shows reduced recruitment of normal MUPs initially but later active dennervation and signs of reinnervation seen. [(21)]
5. Critical illness polyneuropathy: NCS study both sensory and motor shows reduced or absent CMAPs and SNAPs respectively. EMG in early days shows reduced recruitment of MUPs with or without signs of dennervation or reinnervation. Later, as the time advances, MUPs with signs of dennervation and reinnervation may appear. [(22, 23)]

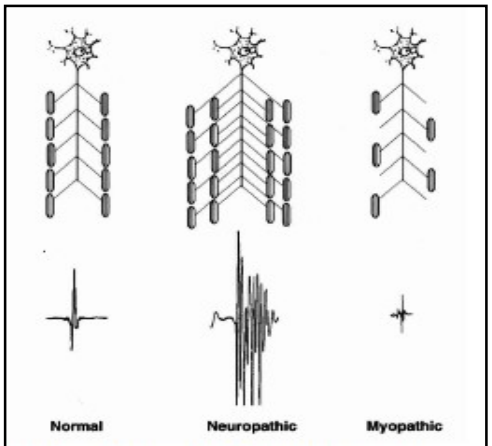
6. Neuromuscular junction disorders including botulism and persistent NMJ blockade: botulism and LEMS exhibits similar EDX picture of low amplitude CMAPs, normal SNAPs, decremental response on slow and incremental response on rapid RNS. MG shows normal CMAPs amplitudes but decremental response on slow RNS that gets repaired on exercise whereas NMJ blockade also shows decremental response that do not repairs after exercise. Single fiber Needle EMG is preferred test to delineate the level of lesion at neuromuscular junction. [24]

7. Critical illness myopathy: Low amplitude CMAPs, normal SNAPs, normal RNS features and EMG shows small, short, polyphasic MUPs with



Temporal dispersion and conduction block in peroneal nerve of a GBS patient

normal or early recruitment. These EMG features suggests primary muscle disease. Long and polyphasic MUPs suggests neurogenic lesion. In adult-onset acid maltase deficiency myogenic MUPs are seen in paraspinal, abdominal or proximal group of muscles. In periodic paralysis, myogenic MUPs may



Morphology of MUPs in normal, neuropathic and myopathic patients: Courtesy- Preston DC, Shapiro BE. Electromyography and neuromuscular disorders

appear later in the course of disease. [23, 25]

8. Miscellaneous causes like phrenic neuropathy can be easily distinguished on EDX studies as it is a localized phenomenon. NCS shows reduced CMAPs in phrenic nerve only. Rest of the motor and sensory nerves remains normal. EMG of diaphragm muscle shows neurogenic MUPs. [12, 26]

Summary

Current review highlights the common neurological issues in ICU that demands EDX evaluation for further management. CIP, GBS and CIM remain major neurological problems that often require EDX evaluation. EDX features to differentiate these three must be well kept in mind while managing patients in ICU. Intensivist should also remember a common protocol for evaluation of these cases. They may ask electromyographer for specific tests to reach the specific lesion site of lower motor neuron involvement. Miscellaneous neurological issues like phrenic neuropathy, neuromuscular junction disorders, and isolated phrenic nerve involvement in motor neuron disease or ALS should also be considered while evaluation.

No conflict of interest

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