INTRODUCTION

Electrodiagnostic (EDX) medicine should be considered an extension of a comprehensive patient

history and physical examination. Combining data found on nerve conduction studies (NCS) and

needle ­ electromyography (EMG), the pathophysiology of a peripheral nerve disease process can be

further defined to illustrate location, duration, severity, and prognosis. It can function as a valuable

aid in patient management, serving as an extension of the clinical exam, but not a substitute.

This chapter focuses on board-related topics about EDX medicine as well as neuromuscular dis-

orders and their associated electrophysiologic changes. It is to be used as a study guide and is not

intended to be an all-inclusive composite. For more elaborate coverage of the subject matter, the reader

is directed to the “References” and “Recommended Reading” sections at the end of this chapter.

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BASIC PERIPHERAL NERVOUS SYSTEM ANATOMY

NEURON ANATOMY AND FUNCTION

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Cell body:

– – – – The cell body (or soma) of a motor or sensory nerve

Cell bodies of motor neurons are located in the anterior (ventral) horn region of the spinal

cord and project an axon distally. It regulates the characteristics of the entire motor unit.

– – Cell bodies of sensory neurons are bipolar cells with two axons (one axon projects proxi-

mally and the other distally) and are found in the dorsal root ganglion (DRG), which is

located outside the spinal cord in the proximity of the intervertebral foramen.

•

Axon:

– – This is the projection from the sensory or motor nerve cell body that propagates current flow

and transports cell nutrition (axonal transport). It can be unmyelinated or myelinated by

Schwann cells.

– – At each spinal level, axons from motor and sensory neurons form ventral and dorsal nerve

roots, respectively, which then combine to become a mixed (sensorimotor) spinal nerve. Each

spinal nerve then branches off to a dorsal and ventral ramus.

– – Motor axons project from their cell bodies to become motor nerve roots.

– Sensory axons project proximally to the spinal cord and distally to become sensory nerve roots.

–

– – Myelin sheaths that cover an axon are electrical insulators that help to accelerate electrical

signal conduction along the axon.

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Nerve:

– A nerve is a bundle of axons that transmit signal to and from various parts of the body.

–

Sensory nerves transmit sensory signals from the body to the central nervous sys-

tem (CNS). Motor nerves transmit motor signals from the CNS to the body’s skeletal

muscles.

– – – – Nerves are covered by nerve connective tissue.

Peripheral nerves:

Motor and sensory nerve fibers combine at various levels in the body (spinal nerve, ventral

ramus, plexus) and ultimately terminate as peripheral nerves.

– – A peripheral motor nerve consists of multiple neural branches from the distal portion of the

axon. They innervate individual muscle fibers.

– The amount of muscle fibers belonging to an axon is the innervation ratio (IR). This ratio

–

­ varies, depending on the function of the motor unit.

– – Muscles of gross movement have a larger amount of their fibers innervated by one axon

(high ratio). Muscles of fine movement have a smaller amount of their fibers innervated by

one axon (low ratio).

nn The axons innervating leg muscles can have a ratio of 600 muscle fibers to one axon

(600:1), while the IR of the eye muscles can be 1 muscle fiber to 1 axon (1:1).

nn The higher the IR, the greater the force generated by that motor unit. A myotome is a

group of muscles that are innervated by one spinal segment.

– – Sensory nerves innervate various segments in the body and are arranged into spinal seg-

mental levels of innervation known as dermatomes.

Neuromuscular junction (NMJ):

– – – – Motor nerves synapse with muscle fibers at sites known as NMJs.

These sites are where the electric impulse propagated along the axon is converted into a

­ chemical reaction. The signal is then translated back into an electrical impulse at the postsyn-

aptic ­ membrane to initiate muscle fiber action potentials (APs).

Muscle fibers:

– – These extrafusal fibers are the final components of the motor unit (see later section on the

Motor Unit). Here, the electrical signal from the postsynaptic NMJ membrane stimulates

muscle fiber depolarization and muscle fiber APs.

– – Muscle fiber characteristics, including twitch response, depend upon the type of alpha motor

neuron by which it is innervated.

Endoneurium

Perineurium

Epineurium

Nerve Connective Tissue (Figure 5–1)

– Endoneurium:

–

nn – Perineurium:

–

nn This is the strong, protective,

connective tissue surround-

ing bundles or fascicles of

­ myelinated and unmyelinated

nerve fibers.

nn It helps strengthen the nerve

and acts as a diffusion barrier.

Individual axons may cross from

one bundle to another along the

course of the nerve.

– Epineurium:

–

nn This is the loose connective

tissue surrounding the entire

nerve that holds the fascicles

together and protects it from

compression.

This is the connective tissue ­ surrounding each individual axon and its myelin sheath.

Myelin sheath

Axon

FIGURE 5–1 Neuronal connective tissue: The internal anatomy

of the nerve.BASIC PERIPHERAL NERVOUS SYSTEM ANATOMY

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The Motor Unit (Figure 5–2)

•

The motor unit is the basic func-

tional element of the neuromuscular

system. It consists of the ­ following

components:

– – Anterior horn cell (motor nerve

cell body)

– – Motor nerve axons

– – Peripheral nerve

– – NMJ

– – Muscle fibers

Plexus

Anterior

horn cell

Spinal

nerve

Nerve root

Peripheral

nerve

Neuromuscular junction

Muscle fiber

Alpha Motor Neurons

•

The three motor neurons listed in

Table 5–1 innervate specific fibers,

FIGURE 5–2 The motor unit.

extrafusal or intrafusal.

•

Needle EMG monitors factors related to the motor unit and thus is limited to evaluating the

alpha motor neurons. The alpha motor neurons and associated motor unit parameters have

been described based on size and physiology (Figure 5–3).

•

The order of recruitment is related to their size, starting with the smaller motor units. This

sequential activation allows for a smooth increase of contractile force and is described by the

Henneman Size Principle.

Henneman Size Principle

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A smaller alpha motor neuron has a lower threshold of excitation, causing it to be recruited

first during voluntary contraction.

•

A larger alpha motor neuron has a higher threshold of excitation and is recruited when more

motor units are needed to generate greater contractile force.

TABLE 5–1 Three Types of Motor Neurons

MUSCLE FIBER TYPES

MOTOR NEURON INNERVATIONS

Alpha Extrafusal fibers—Skeletal muscle

Gamma Intrafusal fibers—Muscle spindle

Beta Intrafusal and extrafusal fibers

FIBER TYPE INNERVATION CHARACTERISTICS

Type I Smaller cell body

Thinner diameter axon

Lower innervation ratio

Slower twitch muscle fibers

Type II Larger cell body

Thicker diameter axon

Higher innervation ratio

Faster twitch muscle fibers334 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

Type II

fiber

Type I

fiber

Myelin

sheath

Type I

motor neuron

Type II

motor neuron

Axon

FIGURE 5–3 Description of Type I and Type II alpha motor neurons.

NERVE FIBER CLASSIFICATION (TABLE 5–2)

•

Nerve fibers vary in their function based on their physiologic characteristics. Their classification

is based on their diameter, conduction velocity (CV), and function.

•

Table 5–2 describes two major classification systems that categorize the different nerve fibers.

•

EDX studies evaluate only Ia (large, myelinated) fibers.

TABLE 5–2 Nerve Fiber Classification

LLOYD

AND HUNT

(SENSORY)

ERLANGER

AND

GASSER

(SENSORY

AND

MOTOR)

DIAMETER

(µM)

VELOCITY

(M/SEC) FUNCTION

Ia fibers A-alpha

fibers

10–20

largest

50–120

fastest

Motor: Alpha motor neurons

Sensory: Muscle spindle

Ib fibers A-alpha

fibers

10–20 50–120 Sensory: Golgi tendon organ, touch, pressure

II fibers A-beta fibers 4–12 25–70 Motor: Intrafusal and extrafusal muscle fibers

Sensory: Muscle spindle, touch, pressure

III fibers A-gamma

fibers

A-delta

fibers

2–8

1–5

10–50

3–30

Motor: Gamma motor neurons, muscle spindle

Sensory: Touch, pain, temperature

IV fibers B-fibers

C-fibers

1–3

<1

3–15

<2

Motor: Postganglionic autonomic fibers

Motor: Preganglionic autonomic fibers

Sensory: Pain, temperatureBASIC PERIPHERAL NERVOUS SYSTEM ANATOMY

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NERVE PHYSIOLOGY

Resting Membrane Potential

•

This is the voltage of the axon’s cell membrane at rest. Normal resting membrane potential

(RMP) is −70 to −90 mV .

•

Leak channels:

– – These are openings in the cell membrane that allow sodium (Na+) and potassium (K+) to

move passively in and out of the cell membrane.

•

Na+–K+ ATP-dependent pumps: (Figure 5–4)

– – A negative potential is maintained inside the cell by actively exporting three Na+ ions while

importing two K+ ions through Na+–K+ ATP-dependent pumps located within the cell’s

­ semipermeable membrane.

– – This keeps each ion against a concentration gradient with a deficit of positive ions inside the

cell. The RMP of the nerve would otherwise dissipate from the ions diffusing through the

ion leak channels.

Cl

Cl

Cl

EXTRACELLULAR

Na+

Cl

Cl

Cl

K+

**+ + + +**

INTRACELLULAR

A-

Na+

Cl

**– – –**

A-

A-

K+

A-

Cl

A-

**–**

Na+/K+

Pump

ATP ADP + Pi

FIGURE 5–4 Na+–K+ ATP–dependent

pump: (Two) K+ ions are imported.

(Three) Na+ ions are exported;

therefore, a negative potential is

maintained inside the cell.

Depolarization

•

When an outside current is applied to a nerve by a stimulator consisting of a cathode (negative

pole) and an anode (positive pole), positive charges on the axon become attracted under the

cathode and lower the membrane potential.

•

The membrane becomes increasingly permeable to Na+, which rushes into the cell through the

opened voltage-gated channels toward an equilibrium. This process of sodium conductance is

the most important event in generating an AP.

•

AP (Figure 5–5):

– – This is a voltage change occurring from an excited cell. The electric impulse propagates

along an axon or muscle membrane. It can also be evoked by a stimulator. The all-or-none

response travels in both directions along the axon.

•

All-or-none response:

– – A stimulus must be strong enough to reach a certain threshold of activation. Once reached,

the AP generated remains at a constant size and configuration.

– – If it is below this threshold, no potential will occur. Any stimulus intensity greater than the

­ threshold will not generate a larger potential.336 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

D

C

B

–

0

+

Outside

Membrane

Inside

+

+

+

+

+

+

Na

– – – – – – – – – –

K

+

+

+

+

+ + + + +

A

–

–

–

–

–

Sodium influx

– – – – –

+

0

Em

–

g

0

–

–

+ + + + + + + + + +

+

+

+

+

+

+

+

+

+

+

–

–

–

–

Potassium efflux

– – – –

+

–

–

–

–

–

–

–

–

–

–

+ + + +

Na

K

Action potential

gK

gNa

**Resting**

**(-90 mV)**

**Activation**

**gate**

FIGURE 5–5 (A) Sodium (gNa) and potassium (gK) ion conductance is depicted over time, resulting in an alteration

of the transmembrane potential and creating an action potential. (B) The spatial relationship of the sodium and

potassium ion influx during an action potential is schematically depicted. Note the alteration of the transmembrane

ionic potential differences corresponding to the depolarization and repolarization. (C) Local circuit currents describe

the pathways of extracellular sodium ions entering the cell and then migrating longitudinally within the cell. (D)

Triphasic extracellular waveform associated with the intracellular monophasic action potential.

•

•

Na+ voltage-gated channels (Figure 5–6):

– – These are protein channels used

for ion exchange. They have acti-

vation and inactivation gates that

undergo conformational changes

from a depolarization.

– – They allow increased Na+ influx

into the cell when activated.

Absolute refractory period:

– – This pertains to the time after

­ closure of the inactivation gates.

They will not immediately reopen.

No AP can be formed at this time,

no matter how strong a repeated

stimulus is used.

– – FIGURE 5–6 Na+ voltage-gated channels.

This may vary in certain disease states (e.g., prolonged in skeletal muscle denervation)

Na+

**OUTSIDE**

**Activated**

**(-90 to +35 mV)**

Na+

**Inactivated**

**(-90 to –35 mV delayed)**

Na+

**Inactivation**

**gate**

**INSIDE**BASIC PERIPHERAL NERVOUS SYSTEM ANATOMY

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•

Relative refractory period:

– – This pertains to the period of time after the absolute refractory period. At this time, an AP

can be elicited with more intense stimulation. This may also be increased or decreased in

certain disease states (e.g., prolonged in skeletal muscle denervation).

Temperature effects (Figure 5–7):

– – The Na+ channels will remain open for approximately 25 μsec. A decrease in temperature

affects the protein configuration and causes a delay in opening and closing of the gates. This

typically changes the waveform appearance, as described later.

– However, the amplitude can drop due to increased temporal dispersion or phase cancellation.

–

– – Also, note the difference in focal cooling compared to generalized limb cooling

Temperature

33°C

(normal)

Onset

latency

(ms)

Peak

latency

(ms)

Negative

spike

duration

(ms)

Amplitude

(µV)

NCV

(ms)

2.1 2.6 1.0 22.0 67

20°C

(focal

cooling)

20°C

(generalized

cooling)

2.4 3.1 1.6 27.5 60

2.8 3.7 1.5 22.0 50

20µV

1ms

FIGURE 5–7 Decreased temperature

effects.

WAVEFORM CHANGES DUE TO A DECREASE IN ­ TEMPERATURE

BELOW 30°C TO 32°C

PARAMETERS CHANGE

Latency Prolonged by 1 msec

Amplitude Increased by 20%

Duration Increased

Conduction velocity Decreased by 10 m/sec

Phases Increased

•

•

Propagation:

– – As Na+ goes into the cell from a depolarization, it moves away from the membrane and

spreads the current down a path of least resistance along the length of the axon. The affinity

to flow back out through the membrane is low due to the myelin sheath covering. Thus, the

potential “jumps” to the next group of Na+ channels, located between the myelin, to areas

called the nodes of Ranvier.

– – This process of propagating a current from one node to another is known as saltatory

conduction.

Directional recording:

Orthodromic Recording

•

•

The action potential is recorded traveling in the direction of its typical physiologic conduction.

Normal physiologic conduction along motor fibers travels away from the spinal cord, whereas

nerve impulses from sensory fibers travel toward the spinal cord.338 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

Antidromic Recording

•

The action potential is recorded traveling in the opposite direction of its typical

­ physiologic conduction.

•

An antidromic motor nerve study records action potential impulses traveling

toward the spinal cord, whereas an antidromic sensory study records sensory

impulses ­ traveling away from the spinal cord.

**Resting**

**(-90 mV)**

**OUTSIDE**

**Slow Activation**

**(-90 to –35 mV delayed)**

K+

K+

**INSIDE**

REPOLARIZATION

•

The process of bringing the depolarized membrane back to

its resting state. It is dependent on Na+ channel inactivation

and K+ channel activation.

•

K+ voltage-gated channels (Figure 5–8):

– – These are protein channels which, after a slight delay,

open from a depolarization. This allows K+ to move out

of the cell to establish a charge equilibrium.

– A delay exists in channel closure, which results in a

–

­ membrane with a hyperpolarized state called an overshoot

phenomenon.

– – This process of potassium conductance eventually returns

the waveform to its baseline due to the K+ leak channels

­ restoring the RMP.

•

•

FIGURE 5–8 K+ voltage-gated channels.

Neuromuscular Junction (Figure 5–9)

The distal portion of a motor axon has small twig-like terminal branches that innervate individ-

ual muscle fibers. This portion of the nerve and single muscle fiber forms the motor endplate.

The axon terminal, containing various neural structures, including mitochondria and synaptic

­ vesicles with acetylcholine (ACh), does not make direct contact with the muscle fiber. Rather, it

remains separate from it by primary and secondary synaptic clefts.

**A**

Axon

Myelin sheath

Teloglial cell Terminal nerve

branches

Myofibrils

Muscle nuclei

**B**

Axon terminal

in synaptic trough

Synaptic vesicles

Subneural

clefts

FIGURE 5–9 The neuromuscular junction. (A) Longitudinal section of the junction. (B) Enlarged view.BASIC PERIPHERAL NERVOUS SYSTEM ANATOMY

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NMJ Components

•

Presynaptic region:

– – This bulbous area at the axon’s terminal zone is comprised of three storage compartments

­ containing ACh. They are contained in packets called quanta consisting of approximately

5,000 to 10,000 molecules.

– – The ACh migrates from the main and mobilization storage compartments to replenish the

­ immediate storage compartment, which is depleted in the process of generating each AP .

This migration of ACh takes approximately 4 to 5 seconds.

STORAGE COMPARTMENTS CONTENT

Main store 300,000 quanta

Mobilized store 10,000 quanta

Immediate store 1,000 quanta

•

•

Synaptic cleft:

– – This is a space 200 to 500 angstroms wide where ACh crosses from the presynaptic region

toward receptors on the postsynaptic region. It contains an enzyme called acetylcholinester-

ase, which degrades ACh into acetate and choline as it crosses the cleft.

Postsynaptic region:

– This is a membrane lined with ACh receptors. It has convolutions to increase its ­ surface

–

area by approximately 10× the surface of the presynaptic membrane. At the crests of

each fold, ­ receptors are located across from the presynaptic active zones, which are the

sites of ACh release. Each ­ postsynaptic ACh receptor requires two molecules of ACh to

become activated.

NMJ Physiology

•

Resting state:

– – During the periods of inactivation, a spontaneous release of a quanta occurs every 5 seconds.

This results in production of one miniature endplate potential (MEPP).

•

Excited state:

– – During the periods of activation, a

nerve depolarization opens voltage-

gated calcium (Ca++) ­ channels. Ca++

floods the nerve terminals and

remains there approximately 200 msec.

– – This leads to the release of multiple

quanta into the synaptic cleft, which

increases the amount of MEPPs. These

MEPPs summate to form an endplate

potential (EPP), which generates a

motor unit action potential (MUAP;

Figure 5–10).

•

Safety factor:

– – The amplitude of an EPP must be high

enough to initiate an AP . Normally,

the EPP’s amplitude is four times

the amount needed to initiate an AP.

However, the EPP’s amplitude drops

each time the EPP is created due to a

drop in immediate available ACh.

**Action**

**potential**

**A X O N T E R M I N A L**

**3**

**Packaging of**

**1**

**ACh in vesicle Acetate**

**4**

**Calcium entry**

**5**

**Vesicle rupture**

**and release of ACh**

**Diffusion**

**of ACh**

**6**

**Combination**

**with receptors**

**Increased permeability**

**to Na+ and K+**

**7**

**Action**

**potential**

**Depolarization**

**(end-plate potential)**

**M U S C L E 2**

**Synthesis**

**of ACh**

**9**

**Uptake of**

**choline**

**8**

**Hydrolysis of ACh**

**(ACh esterase)**

**M E M B R A N E**

FIGURE 5–10 Acetylcholine release and recycling.340 •

•

5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

– – This initial excess amplitude of the EPP is called the safety factor and allows time for ACh to

move from the main and mobilizing storage compartments to replenish the immediate stor-

age compartment. This avoids a drop of the EPP’s amplitude below the threshold needed to

cause an AP. The safety factor depends on two parameters:

nn nn Quantal content: Number of ACh quanta released with each nerve depolarization

Quantal response: Ability of the ACh receptors to respond to the ACh molecules that are

released

Skeletal Muscle Fiber (Figures 5–11 and 5–12)

This is a cylindrical, multinucleated cell containing contractile elements composed of actin and

­ myosin. The sarcomere is a basic unit of a muscle’s myofibril.

A sarcomere (Figure 5–11) runs from Z-line to Z-line. Its size changes during contraction

(Figure 5–12).

Sarcomere

Z Line Z Line

Z Line

Actin filament Myosin filament

Sarcomere

Z Line

H Zone

I Band I Band

A Band

Sarcomere

Z Line Z Line

I Band I Band

H Zone

A Band

**Maximum**

**Contraction Contraction Rest**

I Band

H Zone

A Band

Z Line

I Band

Z Line

I Band I Band

H Zone

Z Line Z Line

I Band I Band

FIGURE 5–11 The sarcomere.

FIGURE 5–12 Sarcomere positional changes.

MUSCLE FIBER CLASSIFICATION (TABLE 5–3)

•

The characteristics of muscle fibers depend on the motor unit by which it is innervated. If a

­ muscle fiber becomes denervated, it will take on the characteristics of the alpha motor neuron

that ­ reinnervates it.

SKELETAL MUSCLE PHYSIOLOGY

•

Muscle fiber contraction (Figure 5–13):

– – An action initiated by muscle fiber depolarization. The stimulus spreads in both directions

on the fiber at 3 to 5 m/sec. It penetrates deeper into the muscle through the T-tubule system.

– This causes Ca++ to be released from the sarcoplasmic reticulum. It binds to the troponin––

tropomyosin

complex and exposes actin’s active sites. Myosin heads, powered by ATP,

bind with the active sites. The actin and myosin filaments slide over each other to shorten

the muscle.

•

Muscle fiber relaxation:

– – Powered by ATP, Ca++ is actively pumped back into the sarcoplasmic reticulum. This allows

the tropomyosin to block actin’s active sites. Absence of ATP results in rigor mortis due to the

actin and myosin filaments remaining permanently joined.PATHOPHYSIOLOGY

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TABLE 5–3 Muscle Fiber Classification

CHARACTERISTICS

TYPE I (SO) SLOW

TWITCH OXIDATIVE

TYPE II-A (FOG) FAST

TWITCH OXIDATIVE-

GLYCOLYTIC

TYPE II-B (FG) FAST

TWITCH GLYCOLYTIC

Alpha motor neuron Small Large Large

Color Dark Dark Pale

Recruitment Early Late Late

Fatigue Highly resistant Resistant Sensitive

Effort Mild (4–8 Hz) Intermediate (20–30 Hz) High (20–30 Hz)

Firing frequency Slow, prolonged Fast, unsustained Fast, unsustained

Movements Fine, precise Gross Gross

Innervation ratio Small Large Large

Amplitude/duration Small Large Large

O2 capacity Aerobic Anaerobic Anaerobic

A. Contraction

B. Relaxation

**C**

**C**

**C**

**C**

**C**

ACh

Action Potential

**MEMBRANE**

**MEMBRANE**

Sarcoplasmic reticulum

(Calcium pump)

Sarcoplasmic reticulum

(Calcium pump)

**C**

**C C**

**C**

**C C**

**C**

**C**

**C**

**C**

**C**

**C C C**

**C**

**C**

**C C**

**C**

Tropomyosin/

Troponin

**C**

**C**

**C C**

**C C**

**C**

**C**

**C**

Pi

ADP

ADP

**C**

**C**

Pi

ATP

ATP

Actin

**MYOSIN FILAMENT MYOSIN FILAMENT**

FIGURE 5–13 Muscle contraction: Excitation–contraction coupling in the muscle. This shows an action potential that causes

the release of calcium ions from the sarcoplasmic reticulum and then reuptake of the calcium ions by a calcium pump.

n

PATHOPHYSIOLOGY

DEMYELINATION INJURY (FIGURE 5–14)

•

This is an injury to the myelin sheath of the

nerve, but the axon remains intact. Demyelination

increases the membrane capacitance due to the

loss of myelin insulation, thus hindering saltatory

conduction.

•

This translates to slower signal conduction along

the axon. The trophic factors of the nerve are

­ maintained, and myelin regeneration is pos-

sible due to Schwann cell proliferation. Acutely,

­ conduction block can occur. With time, remy-

elination can occur. In some chronic disease

states, demyelination and remyelination occur

repeatedly.

A B

FIGURE 5–14 Demyelination. (A) Normal nerve.

(B) Injured segment with myelin breakdown.342 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

Conduction Block

Failure of an AP to propagate past an area of demyelination along axons that are ­ otherwise struc-

turally intact is known as conduction block. It can present as a >50% drop in CMAP amplitude

between proximal and distal stimulation sites across the area of injury.

AP, action potential; CMAP, compound muscle action potential.

•

Etiologies:

– Focal compression causing a transient ischemic episode, edema, or myelin invaginations

–

with paranodal intussusceptions (Figure 5–15).

– – Chronic diseases causing degradation of myelin leading to peripheral neuropathies

Basement membrane Schwann cell “Pseudo-node”

**Axon**

Myelin

NCS EMG

Latency: Prolonged

Conduction velocity: Decreased

Temporal dispersion: Increased

Amplitude: May decrease secondary to ­ temporal dis-

persion and phase cancellation

Normal insertional activity

Resting activity: Normal, ± myokymia

Recruitment: Normal or decreased

MUAP: Normal

Nodal end

loops of myelin

True node

Schwann cell

processes

FIGURE 5–15 Paranodal intussusception. Diagram of an invaginating paranode into an adjacent one.

•

EDX findings of demyelination:

– – Because demyelination affects the speed of signal conduction along a nerve, it can affect

measurements related to time on NCSs, notably latency, CV, and temporal dispersion.

EMG, electromyography; MUAP, motor unit action potential; NCS, nerve conduction study.

•

Recovery:

– – Self-limited:

nn The pathology can reverse with cessation of the insult-

ing event. Transient ischemia can be immediately

reversed, but edema can take several weeks.

– – Remyelination (Figure 5–16):

nn This is a process of repair in which the demyelinated

region develops new myelin produced by the Schwann

cells. This new myelin is thinner with shorter inter-

nodal distances. CV improves but is usually slower

than normal.

AXONAL INJURY (FIGURE 5–17

AND FIGURE 5–18)

•

An injury to the axon may present in one of two typical

forms: axonal degeneration or Wallerian degeneration.

Both of these can affect the cell body and cause a central

chromatolysis.

A B C

FIGURE 5–16 Remyelination.

(A) Myelin digestion and Schwann cell

proliferation. (B) Myelin is removed.

(C) Remyelination is complete.PATHOPHYSIOLOGY

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I

II

III

IVb

IVa

FIGURE 5–17 Axonal injury. (I) Normal nerve cell. (II) Postinjury: Nissl substance degenerates. (III) Swollen cell body

with eccentric nucleus. (IVa) Cell death. (IVb) Cell recovery.

Nerve cell body

Nucleus

Axon

Internode

Node of Ranvier

Schwann cell

Nucleus

Motor endplate

Muscle

Normal

Wallerian

degeneration

Axonal

degeneration

FIGURE 5–18 Schematic representation of axonal injuries.

•

•

•

•

Axonal degeneration (Figure 5–18):

– A nerve injury that begins in a “dying back” fashion and affects the nerve in a length-depen-

–

dent manner. Degeneration of the axon starts distally and ascends proximally.

Wallerian degeneration (Figure 5–18):

– – At the site of a nerve lesion, the axon degenerates distally. The nerve segment proximal to

the injury site is essentially intact with some minor dying back at the lesion site 1 to 2 cm.

– – For the distal motor axons, the degeneration is generally complete in 7 days.

– – For the distal sensory axons, the degeneration is generally complete in 11 days.

Etiology:

– – Pathology can occur from: (a) focal crush, (b) stretch, (c) transection, or (d) peripheral

neuropathies.

EDX findings:

– – Axonal injuries affect the amplitude on nerve conduction waveform, as it is representative of

the fastest axons from a nerve. A decrease in amplitude would represent axonal loss.344 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

NCS EMG

Amplitude: Decreased

Temporal dispersion: Normal

Conduction velocity and distal latency: Mild slowing of

both may occur if the largest and fast conducting axons

are lost

Insertional activity: Abnormal

Resting activity: Abnormal

Recruitment: Decreased

MUAP: Abnormal

EMG, electromyography; MUAP, motor unit action potential; NCS, nerve conduction study.

•

Recovery:

– Collateral sprouting (Figure 5–19):

–

This is a process of repair in which a neurite sprouts off the axon of an intact motor unit

and innervates denervated muscle fibers of an injured motor unit. The sprouts con-

nect with smaller terminal branches, thinner myelin, and weaker NMJs compared to

an uninjured axon. Increased fiber type grouping occurs as muscle fibers become part

of the new motor unit and take on its characteristics, increasing the size of its terri-

tory. This remodeling results in motor units with poor firing synchronicity, secondary

to the immature terminal sprouts. This results in polyphasic ­ waveforms with increased

amplitudes.

– Axonal regrowth (Figure 5–20):

–

This is a process of repair in which the axon will regrow down its original pathway toward

its muscle fibers. It will travel approximately 1 mm/d or 1 in./mo (35 mm/mo) if the ­ supporting

­ connective tissue remains intact. These axons will have a decreased diameter, thinner

myelin, and shorter ­ internodal distance. With reinnervation, low-amplitude, long-duration,

and ­ polyphasic potentials known as nascent potentials are formed. If the connective tissue is

not intact to guide proper nerve regrowth, a neuroma can form with failure to reach the final

end organ. Concomitantly, the shorter the distance from injury to end organ, the higher the

likelihood for a better prognosis.

A

Normal

600 µV

B

2-3 weeks

post-den

600 µV

C

1-2 months

post-den

1,200 µV

D

2-6 months

post-reinn

7,000 µV

E

6 mo – 2 yr

post-reinn

6,000 µV

F

Total denervation

100 µV

FIGURE 5–19 Motor unit remodeling. (A) Type I:

Light circles; Type II: Dark circles. Depolarization

of one of the motor units results in a 600 mV

MUAP. (B) Following degeneration of the Type II

MU at 2 to 3 weeks, the Type I MUAP still yields

a 600 mV potential. (C) Within 1 to 2 months,

the Type II muscle fibers have atrophied, and

collateral sprouting from Type I fibers is beginning

to reinnervate them. The Type I motor unit territory

has subsequently collapsed due to Type II fiber

atrophy, causing a larger MUAP (1,200 mV). (D) As

the connections mature, the MUAP demonstrates

a further increase in amplitude (7,000 mV) and

number of phases. By 6 months, all muscle

fibers belonging to the Type I motor units are

of the same fiber type; i.e., Type II fibers have

been converted to Type I fibers. (E) As maturity

continues, the MUAP may decrease its amplitude

and phases due to the collaterals conducting

potentials more rapidly. (F) An example of

complete denervation.

MUAP, motor unit action potential.

Source: Copyright ©1995 American Association of

Electrodiagnostic Medicine.PATHOPHYSIOLOGY

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Internodal

distance

MYELIN

Internodal

distance

CHARACTERISTICS NEUROPRAXIA AXONOTMESIS NEUROTMESIS

Etiology Nerve compression injury Nerve crush injury Nerve transection injury

Description Axon is intact Local

myelin injury

Conduction block

Axonal interruption

Connective tissue/

Schwann cell intact

Conduction failure

Axonal interruption

Connective tissue

disruption

Conduction failure

Nerve conduction studies The signal is normal

with stimulation distal to

the lesion but abnormal

with stimulation proxi-

mal to it

Conduction resembles

neuropraxia for 4–5 days,

until Wallerian degenera-

tion occurs

Conduction initially

­ resembles axonotmesis

but does not demonstrate

recovery

Stimulation

proximal

to lesion

Recording electrodes

on palmar surface over

hypothenar eminence

Stimulation

distal to

lesion

Lesion

Waveform

distal to

lesion:

Immediate

2 weeks

>2 weeks

Waveform

proximal

to lesion:

\_\_\_\_\_\_

\_\_\_\_\_\_

Waveform

distal to

lesion:

Immediate

2 weeks

\_\_\_\_\_\_

weeks–

months

Waveform

proximal

to lesion:

\_\_\_\_\_\_

\_\_\_\_\_\_

Waveform

distal to

lesion:

Immediate

2 weeks

\_\_\_\_\_\_

2 years

\_\_\_\_\_\_

Waveform

proximal

to lesion:

\_\_\_\_\_\_

\_\_\_\_\_\_

\_\_\_\_\_\_

EMG Normal/decreased

recruitment

Abnormal activity Abnormal activity

**AXON**

FIGURE 5–20 Axonal regrowth: Axonal diameter is decreased. Myelin is thinner. Internodal distance is shorter.

Collateral Sprouting Versus Axonal Regrowth

If an axon regrows to innervate its original muscle fibers, but collateral sprouting to these fibers

has occurred, the nerves possessing the largest axon, thickest myelin, and strongest NMJ will

prevail and keep the muscle fibers.

NMJ, neuromuscular junction.

NERVE INJURY CLASSIFICATION (TABLES 5–4 AND 5–5)

•

Two classification systems categorizing nerve injuries are:

– – Seddon classification (Table 5–4)

– – Sunderland classification (Table 5–5 and Figure 5–21)

TABLE 5–4 Seddon Classification

EMG, electromyography.346 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

TABLE 5–5 Sunderland Classification

TYPE 1 Conduction block (neuropraxia)

TYPE 2 Axonal injury (axonotmesis)

TYPE 3 Type 2 + Endoneurium injury

TYPE 4 Type 3 + Perineurium injury

TYPE 5 Type 4 + Epineurium injury (neurotmesis)

**Type 1**

Perineurium

Endoneurium

Axon with

complex sheath

Epineurium

**Type 2**

**Type 3**

**Type 4**

**Type 5**

FIGURE 5–21 Sunderland classification.

n

CLINICAL INSTRUMENTATION

Electrodiagnostic studies consists of NCSs and needle EMG.

ELECTRONIC CIRCUITRY (OHM’S LAW)

•

An electric current passes through a wire at an intensity of the current (I) measured in amperes,

equal to the voltage (V) from an electromotor source measured in volts divided by the resis-

tance (R) measured in ohms. The following formula is known as Ohm’s Law:

Current = Voltage/Resistance (I = V/R or V = I × R).

ELECTRODIAGNOSTIC INSTRUMENTATION (FIGURE 5–22)

ELECTRODES

These devices are used to record or stimulate the skin surface, muscle, or nerve. An electrode can

be an active, reference, stimulating, or ground electrode. They come as either surface or needle elec-

trodes. To obtain a proper reading, the impedance (resistance) between the electrode and skin must

be kept low by removing skin lotions, oils, gels, and so on.CLINICAL INSTRUMENTATION

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**A**

Analog

display Memory

PATIENT

G1

Active

G2

Reference

Ground

**B**

Variable

gain

Differential

amplifier

**C**

High and low

frequency

filters

**D**

Analogic

digital

conversion

Microprocessor

**E**

**Video monitor**

**Cathode ray tube**

Audio

monitor

Nerve stimulator

**F**

FIGURE 5–22 Electrodiagnostic instrumentation. (A) A patient with recording electrodes has a peripheral nerve

excited with a stimulator (F). (B) The differential amplifier receives the action potential. (C) The signal is filtered.

(D) The analog signal is converted to a digital representation while being fed to a loudspeaker. (E) The signal is

displayed on a cathode ray tube. (F) Stimulator is used to excite the peripheral nervous system.

•

Recording electrodes:

These are devices placed on the skin or in the soft tissue to pick up electrical activity from the

muscle or nerve. See next section on surface vs. needle electrodes for further details.

– Active electrode (G1):

–

This pickup records the electrical activity from a nerve AP. In a sensory nerve action

potential (SNAP), the recording electrode is placed directly over the nerve, and the

electrical activity from the nerve is recorded. The recording electrode for a motor nerve

study (compound muscle action potential [CMAP]) is placed over the motor endplate of

a muscle that is innervated by that nerve. The CMAP that is recorded represents the sum-

mation of electrical activity generated by muscle fibers; it is an indirect representation of

electrical activity generated by a motor nerve.

– Reference electrode (G2):

–

This pickup is placed over an electrically neutral area (tendon or bone) during a sensory or

motor nerve study.

•

•

Surface electrodes (Figure 5–23):

Surface electrodes are placed on the

skin to record nerve or muscle APs.

They are typically either metal elec-

trodes or disposable electrode stickers

lined with an adhesive backing and

conductive gel.

Needle electrodes:

These electrodes are inserted

through the skin to record muscle or

nerve APs. If used for NCS, the wave-

form’s amplitude and CV cannot be

assessed because the needle samples

only a few fibers.

– Monopolar needle electrode

–

(Figure 5–24):

This is a 22- to 30-gauge Teflon-

coated needle with an exposed tip

of 0.15 to 0.2 mm2

.

Disposable ground

electrode

Disposable strip

electrode

Standard bar

electrode

Flat disc

electrode (1 cm)

Wire ring

electrode

Circular ground

electrode

FIGURE 5–23 Various types of surface electrodes.348 •

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5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

Advantages:

nn Inexpensive

nn Conical tip: Omni-directional recording

nn Less painful (Teflon decreases friction)

nn Larger recording area (2× concentric)

nn Records more positive sharp waves (PSWs) and more abnormal

­ activity in general

Disadvantages:

nn Requires a separate needle or surface reference

nn Nonstandardized tip area

nn Teflon fraying

nn May have more interference if the reference is not near the record-

ing electrode

– Standard concentric (Coaxial) needle electrode (Figure 5–25):

–

This is a 24- to 26-gauge needle (reference) with a bare inner wire (active).

Advantages:

nn Standardized exposed area

nn Fixed location from reference

nn Less interference

nn No separate reference

nn Used for quantitative EMG

Disadvantages:

nn Beveled tip: Unidirectional recording

nn Smaller recording area

nn MUAPs have smaller amplitudes

nn More painful

– Bipolar concentric needle electrode (Figure 5–26):

–

This is a needle with the active and reference wires within its lumen.

Advantages:

nn Best for isolating MUAP

nn Less artifact

Disadvantages:

nn Expensive

nn More painful

– Single-fiber needle electrode (Figure 5–27):

–

This is a needle (reference) consisting of an exposed 25-μm diameter

wire (active).

Advantages:

nn Looks at individual muscle fibers

nn Used to assess fiber type density

nn Used to assess jitter

nn Used to assess fiber blocking

nn Helpful in assessing NMJ disorders and motor neuron disorders

Disadvantages:

nn Not used for traditional EMG

nn Expensive

Ground electrode:

This is a zero-voltage, surface reference point placed between the

­ recording electrode and the stimulating electrode.

Stimulating electrode (Figure 5–28):

This is a bipolar device used to apply an electrical impulse to a nerve to

initiate a nerve AP. The stimulator has a cathode and an anode pole:

– The cathode terminal generates a negative impulse that attracts positive

–

charges from the axon.

– The anode terminal generates a positive impulse that attracts negative

–

charges from the axon.

E-1

E-2

R

FIGURE 5–24 Monopolar

needle electrode.

E-1

E-2

R

FIGURE 5–25 Concentric

needle electrode.

E-1

E-2

R

FIGURE 5–26 Bipolar

needle electrode.

E-1

E-2

R

FIGURE 5–27 Single-

fiber electrode.CLINICAL INSTRUMENTATION

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Anodal Block

A theoretical local block that occurs when reversing the stimu-

lator’s cathode and anode. This hyperpolarizes the nerve, thus

inhibiting the production of an action potential.

FIGURE 5–28 Bipolar stimulator.

NERVE CONDUCTION STIMULATION

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NCSs are performed by electrically stimulating the nerve and recording the signal. The

recorded signal is affected by multiple technical factors, including stimulation intensity and

duration as well as noise and interference signal.

Stimulation Intensity

•

Threshold stimulus:

– – This is an electrical stimulus occurring at an intensity level just sufficient enough to produce

a detectable evoked potential from the nerve.

•

Maximal stimulus:

– – This is an electrical stimulus at an intensity level in which no further increase in an evoked

­ potential will occur from the nerve with added stimulus intensity.

•

Submaximal stimulus:

– – This is an electrical stimulus at an intensity below the maximal stimulus level but above

the threshold level. This can lead to a falsely lower recorded amplitude and prolonged

latency ­ reading, which can give the false impression of an axonopathy or conduction

block.

•

Supramaximal stimulus:

– – This is an electrical stimulus at an intensity at least 20% above the maximal stimulus and is

­ typically used for NCS.

– With stimulus intensity set too high, unwanted results may occur due to volume conduc-

–

tion. Volume conduction occurs when the stimulus current spreads through tissue surround-

ing the nerve. Skin, extracellular fluid, muscles, and other nerves may be stimulated, which

can lead to:

nn Decreased conduction times and shortened latencies

nn Altered waveforms

nn Amplitudes remain unchanged

Stimulation Duration

Usually stimulus duration is set at 0.1 msec and may be increased incrementally to ensure supra-

maximal stimulation. If a monopolar needle is used for stimulation, start at 0.5 msec. Longer stimu-

lus duration will cause more pain.

Stimulation Averaging

This process extracts the desired neurophysiologic signal from larger noise and interference signals.

These unwanted signals can occur from biological or environmental sources, such as EMG audio

­ feedback, needle artifact, 60 hertz (Hz) line interference, preamplifier proximity to the machine,

fluorescent lights, or the patient.

•

Signal-to-noise ratio (S:N):

The process of averaging improves the S:N by a factor that is the square root of the number of

­ averages performed. The number of averages must be increased by a factor of four to double

the S:N.

S : N

=

´

Signalamplitude # of averages performed

Noiseamplitude

.350 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

Stimulus Artifact

This is a defect seen at the time the stimulus is applied to the skin and represents current spread to

the electrode. It can be minimized by:

nn Placing the ground electrode between the recording electrode and stimulator

nn Appropriate anode and cathode placement

nn Cleansing the skin from dirt, perspiration, and lotions

DIFFERENTIAL AMPLIFIER (FIGURES 5–22B AND 5–29)

This is a device within a preamplifier that responds to alternating currents of electricity. It cancels

waveforms recorded at both the active and reference pickups and amplifies the remaining poten-

tials (Figure 5–29). It should have a high impedance and common mode rejection but low noise

from within the system.

Common Mode Rejection Ratio

This refers to selectively amplifying different signals and rejecting common ones. It is usually

expressed as dB and should be ≥90 dB. The larger the CMRR, the more ­ efficient the amplifier.

dB, decibels; CMRR, common mode rejection ratio.

G1

Active

Differential signal =

Active – reference

60 Hz interference

Variable

gain

Differential

amplifier

G2

Reference

FIGURE 5–29 Schematic representation of

differential amplifier function. A differential

amplifier only amplifies the difference in the signal

present at the active and reference inputs. When

60 Hz interference is the same at both inputs, it

is eliminated, leaving only the difference signal,

which is the action potential, being measured.

FILTERS (FIGURE 5–30)

This device, composed of resistors and capacitors, functions to exclude unwanted waveforms from

being recorded.

•

Types of filters:

– High-frequency (low pass) filter (HFF):

–

An HFF removes signals with frequencies

higher than its cutoff setting. Signals with fre-

quencies lower than (below) the cutoff setting

are not affected. This affects the faster portions

of the summated waveform.

– Low-frequency (high pass) filter (LFF):

–

An LFF removes signals with frequencies lower

than its cutoff setting. Signals higher than (above)

the cutoff setting are not affected. This affects the

slower portions of the summated waveform.

•

Filter settings:

– – Sensory NCS: 20 Hz to 10 kHz

– – Motor NCS: 2 Hz to 10 kHz

– – EMG: 20 Hz to 10 kHz

•

Filter adjustments:

– Changes in waveforms can be expected with

–

increasing the LFF (e.g., increase from 1 to 500 Hz)

or lowering the HFF (e.g., decreasing from 10,000

to 500 Hz, while maintaining the LFF at 1 Hz).

Band width viewed

by instrument

I

II

0

¥

Frequency

FIGURE 5–30 The frequency bandwidth. This is a

schematic representation of the frequencies the

filters have allowed the instrument to view. (I):

Low-frequency filter. (II): High-frequency filter.CLINICAL INSTRUMENTATION

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Effects of Filter Changes on NCS Waveforms

Elevating the Low-Frequency Filter

(Figure 5–31 I–IV)

•

•

•

•

Reduces the peak latency

Reduces the amplitude

Changes potentials from bi- to triphasic

Does not change the onset latency

Reducing the High-Frequency Filter

(Figure 5–32 I–IV)

•

•

•

•

Prolongs the peak latency

Reduces amplitude

Creates a longer negative spike

Prolongs the onset latency

SENSORY MOTOR EMG

Sweep Speed 5 msec 2 msec 10 msec

Sensitivity 10 µ V 5 mV 100 µ V (Insertional activity)

1 mV (Recruitment pattern analysis)

FIGURE 5–31 Elevating the low-frequency filter:

Sequential elevation of low-frequency filters

(I–IV) from 1 to 500 Hz.

FIGURE 5–32 Reducing the high-frequency filter:

Sequential reduction of high-frequency filters (I–IV)

from 10,000 to 500 Hz.

SCREEN

•

Once a signal has been recorded, amplified, filtered, and passed through, the analog-to-digital

­ converter is displayed on the computer screen. A grid is projected on the screen with the hori-

zontal axis representing sweep speed and the vertical axis representing sensitivity. Each of these

parameters can be adjusted to manipulate the recorded waveform for an accurate measurement.

•

•

Sweep speed pertains to the time allocated for each x-axis division and is measured in milliseconds.

Sensitivity pertains to the height allocated for each y-axis division and is measured in millivolts

(mV) or microvolts (μV). The term gain is sometimes used interchangeably with sensitivity. Gain is

actually a ratio measurement of output to input and does not have a unit value such as mV or μV.

•

Settings:

SAFETY ISSUES

•

Each aspect of the electrodiagnostic (EDX) exam has certain risk factors. During NCS, electri-

cal risks need to be ­ considered; in needle EMG, certain bleeding risks should be addressed.

Though there are no absolute contraindications, these relative risks are weighed against com-

mon sense, and data obtained in the history (e.g., unexplained bleeding or ecchymosis, cardiac

pacemakers, or defibrillators).

•

Electrical risk factors:

– – Exercise caution in routine EDX studies regarding applying a current to the body.

Theoretically, delivering a stimulus may affect factors of cardiac conduction or cause bodily

injury from ­ electrical shock.

•

Cardiovascular devices:

– – Far-field potential generated by routine NCS does not cause electrical activity that would

­ create a detectable stimulation. They pose no risk to implantable pacemakers or intracardiac352 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

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­ defibrillators. Yet, a 15 cm (6 inch) separation is suggested between the stimulator and

any wires, ­ intravenous (IV) lines, or catheters as a general rule. In addition, one should

avoid stimulating the brachial plexus on the same side as a pacemaker or internal cardiac

defibrillator.

Contraindications:

– – External cardiac pacemakers: External pacing wires can be electrically sensitive to NCS

stimulations.

– – Central line catheters may pose a risk of generating a stimulus in the heart. However,

peripheral IV lines are not considered to be problematic.

Bleeding risks:

– – Clinically relevant bleeding issues from an EMG are extremely rare. Considerations to alter

the EMG are based on physician comfort for patients taking antiplatelet or anticoagulant

medications or with coagulopathies.

– – It is not routinely encouraged to hold anticoagulant or antiplatelet medications for this

study. Caution may be exercised in patients with platelet counts <50,000 per microliter and

the International Normalized Ratio (INR) has been suggested to be acceptable at 3.0.

Other:

– – – – – – Studies at the neck need to take into account the location of the carotid sinus and vagus

nerve. Stimulating these could affect the rhythm of the heart.

Electrodes should not be placed in a manner where they read a response across the heart.

Other common sense issues should be taken into account to avoid external influences on the

NCS, such as avoiding contact with exposed metal or wet surfaces.

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NERVE CONDUCTION STUDIES

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These studies assess the ability of peripheral nerves to conduct electrical impulses. A represen-

tative waveform is generated by nerve stimulation, and its parameters are evaluated to monitor

peripheral neuronal function.

Waveforms:

– – These recorded potentials represent a com-

pilation of multiple sinusoidal waves. The

different phases and amplitudes of each

waveform summate or cancel to create a

final potential.

– – The final potential displayed represents an

average of all the subcomponent’s fre-

quencies (Figure 5–33).

– – Frequency is defined as the number of

times the same event occurs in 1 second

(cycles per second) and is measured in Hz.

(1, 0, 1)

(3, 0, 0.33)

(5, 0, 0.2)

(7, 0, 0.14)

(9, 0, 0.11)

A B

(0.5, 90, 2)

(2, 0, 0.7)

(3, 0, 0.5)

(4, 0, 0.3)

(5, 0, 0.2)

PARAMETERS (FIGURE 5–34)

•

Latency (Table 5–6):

– Onset latency is the time required for an

–

electrical stimulus to initiate an evoked

potential.

nn For sensory studies, onset latencies

reflect conduction along the fastest

fibers.

nn For motor studies, distal latencies

­ represent the time interval from

the motor stimulation site to the

NMJ, time delay across the NMJ,

FIGURE 5–33 Waveform subcomponents (summation).

(A) A square wave and five subcomponent sine waves,

which, when added together, result in the square

wave. (B) A more biologic appearing potential and

its subcomponent sine waves. The frequency, phase

shift, and relative amplitude are described later each

subcomponent waveform.NERVE CONDUCTION STUDIES

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NERVE FIBER NERVE RESPONSE

Motor Latency of activation: This is the time between the initiation of the electrical stimulus and the

beginning of saltatory conduction. It is typically 0.1 Conduction: This is the saltatory conduction of an AP along the myelinated axons to its termi-

nal branches, unmyelinated twigs, and NMJ.

Synaptic transmission: This is the chemical transmission of the signal across the NMJ to

initiate a single-fiber AP. It takes 0.2–1 msec.

Sensory Latency of activation: Same process as earlier

Conduction: Same process as earlier

Synaptic transmission: This does not apply due to absence of an NMJ.

and ­ depolarization of the muscle

fibers. Conduction along the fastest

fibers does not correlate with onset

latency.

– Peak latency represents the latency

–

along the majority of the axons and is

measured at the peak of the waveform

amplitude. Both latencies are primar-

ily dependent on the myelination of a

nerve.

Duration

Distal latency

Baseline

to peak

amplitude

Peak latency

Peak

to peak

ampli-

tude

•

Conduction velocity:

– – This is the speed an impulse travels

along a nerve and is primarily depen-

dent on the integrity of the myelin

FIGURE 5–34 Waveform parameters.

TABLE 5–6 Nerve Responses of Motor and Sensory Nerve Fibers

msec or less.

AP, action potential; NMJ, neuromuscular junction.

sheath. It is calculated by dividing the change in distance (proximal stimulation site in mil-

limeter minus distal stimulation site in millimeter) by the change in time (proximal latency

in milliseconds minus distal latency in milliseconds).

– Normal values are generally >50 m/sec in the upper limbs and >40 m/sec in the lower limbs.

–

– – It can be decreased with nerve injury and from technical factors. It should remain normal

even in severe axonal injuries, as NCSs record the velocity of fastest surviving nerve fibers.

CV VARIATIONS:

Age

•

•

•

CV for a newborn is 50% that of an adult. At 1 year, it is 80% that of an adult. It is equal to an

adult by 3–5 years.

Due to segmental demyelination/remyelination and large fiber loss associated with normal

aging, typical changes can be seen.

After the fifth decade, the CV decreases 1–2 m/sec per decade.

CV, conduction velocity.

MEASUREMENTS DESCRIPTION

Onset latency • This represents initiation of a conduction response along the fastest axons.

•

It is recorded at the initial deflection from baseline.

Peak latency • This represents initiation of conduction along the majority of the axons.

− Is a more accurate measure of slowing

−

•

It is recorded at the peak of the waveform response.354 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

Temperature

•

Normal is approximately 32°C for the upper limbs and 30°C for the lower limbs.

•

It decreases 2.4 m/sec per 1°C dropped.

•

A 5% decrease in CV has been described for each 1°C drop below 29°C.

CV, conduction velocity.

•

•

•

•

Amplitude:

– – – – This is the maximum voltage difference between two points.

In sensory studies, the sensory nerve amplitude reflects the sum of activated sensory nerve

fibers and their synchronicity of firing. It is commonly measured from baseline to negative

peak or first negative peak to the next positive peak.

– In motor studies, the amplitude reflects the number of muscle fibers that have been activated.

–

Recordings are typically measured from baseline to the negative peak. While most cases of

reduced CMAP amplitudes are due to a loss of axons (as in a typical axonal neuropathy), other

causes of low CMAP amplitude include conduction block, some NMJ disorders, and myopathies.

Duration:

– – This is measured from the initial deflection from baseline to the first baseline crossing.

– – In sensory nerves, it is a measure of synchrony of the sensory nerve fibers firing.

– – In motor nerves, it is a measure of synchrony of the individual muscle fibers firing.

Area:

– – This is a function of both the amplitude and duration of the waveform.

Temporal dispersion (Figure 5–35):

– – This reflects the range of conduction velocities of the fastest and slowest nerve fibers. The

waveform spreads out (disperses) with proximal compared to distal stimulation. The area

under the waveform remains essentially constant.

– This is due to slower fiber conduction reaching the recording electrode later than faster fibers.

–

– – This is not usually seen with more distal stimulation when slow and fast fibers reach the

­ recording electrode at relatively the same time.

x 2x

3x

**A B C D**

I

II

III

Fast

Medium

Slow

FIGURE 5–35 Temporal dispersion. Three axons of

various conduction speed. (I) Fast conduction axon.

(II) Medium conduction axon. (III) Slow conducting

axon. The signal is measured at different points along

the nerve at site A, B, C; then conduction begins

at the left and proceeds to the right. At point A, the

signal of each axon arrives almost simultaneously,

producing a very compact recorded response. At

point B, the signals are less well synchronized,

producing a smaller amplitude and longer duration

response, and this spreading is increased by the time

the signals arrive at point C and point D.

•

Phase cancellation: (Figures 5–36 and 5–37):

– – When comparing a proximal to distal stimulation, a drop in amplitude and increase in dura-

tion occurs, most notably with a SNAP because of its short duration.

– – When the nerve is stimulated, the APs of one axon may be out of phase with neighboring

ones. The negative deflections of one axon can then cancel the positive deflection of another,

reducing the amplitude. The summation of these axons creates an AP that appears as one

long prolonged wave.

– – – – For this reason, a drop of 50% is considered normal when recording a proximal SNAP.

The CMAP does not have as much of a drop in amplitude because it has a longer duration

­ waveform, and also because of NMJ cushioning. Thus, a smaller decrease in amplitude of

­ approximately 15% is expected.NERVE CONDUCTION STUDIES

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SENSORY NERVE ACTION POTENTIALS (SNAP)

•

A sensory nerve study represents the conduction of an impulse along the sensory nerve fibers.

It can also be useful in localizing a lesion in relation to the DRG (Figure 5–38).

•

The DRG is located in the intervertebral foramen and contains the sensory cell body. Lesions

proximal to it (injuries to the sensory nerve root or to the spinal cord) preserve the SNAP

waveform despite clinical sensory abnormalities. This is because axonal transport from the cell

body to the peripheral axon continues to remain intact. SNAPs are typically considered more

­ sensitive than CMAPs in the detection of an incomplete peripheral nerve injury.

**Stimulation of Nerves Distally**

**Individual**

**responses**

**Summated**

**responses**

Fast conducting nerve

Slow conducting nerve

**Stimulation of Nerves Proximally**

**Individual**

**responses**

**Summated**

**responses**

Fast conducting nerve

Slow conducting nerve

FIGURE 5–36 Sensory—SNAP phase

cancellation. Open arrows indicate

stimulation of the nerve distally; the

phases from the individual SNAPs

summate. Closed arrows indicate

stimulation of the nerve proximally;

with the increased distance, the

phases separate enough by the time

they reach the recording electrodes to

summate less or even cancel.

SNAP, sensory nerve action potential.

**Stimulation of Nerves Distally**

**Individual responses Summated responses**

Fast conducting nerve

Slow conducting nerve

**Stimulation of Nerves Proximally**

**Individual responses Summated responses**

Fast conducting nerve

Slow conducting nerve

FIGURE 5–37 Motor—CMAP phase cancellation. Open arrows indicate stimulation of the nerve distally resulting in

the discharge of two MUAPs that produce a potential with twice the size. Closed arrows indicate stimulation of the

nerve proximally, resulting in two MUAPs that still summate in phase because of the long duration of the MUAPs’

negative phases.

CMAP, compound motor action potential; MUAP, motor unit action potential.356 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

**Postganglionic lesion**

**DRG**

**Preganglionic lesion**

**DRG**

**DRG**

**Normal Axons**

**Degenerated Axons**

**Dorsal Root Ganglion**

FIGURE 5–38 Postganglionic injury results in Wallerian

degeneration of both motor and sensory axons. There is

physical separation of the axon from the cell bodies in the

DRG and the ventral portion of the spinal cord. Compound

motor action potential and SNAP responses are diminished

or absent. Preganglionic injury produces the same injury

to the motor fibers but allows the peripheral sensory fibers to

remain in contact with their cell body. As a result, SNAPs are

normal in this injury.

DRG, dorsal root ganglion; SNAP, sensory nerve action

potential.

•

Technical considerations:

– Antidromic studies:

–

nn Are easier to record a response than orthodromic

studies

nn May be more comfortable than orthodromic

studies due to less stimulation intensity

required

nn May have larger amplitudes due to the nerve

being more superficial at the distal recording sites

– Recording electrodes:

–

nn The active and reference pickup should be at least

4 cm apart. Less than this distance will alter the

waveform in the following manner (Figure 5–39).

Results When Electrode

Separations <4 cm Apart

PARAMETERS CHANGE

Peak latency Decreased

Amplitude Decreased

Duration Decreased

Rise time Decreased

50 µ*V*

2 ms

V

**Trace**

**Interelectrode**

**Distance**

**(***cm***)**

**Amplitude**

**(**µ*V***)**

**Latency**

**(***ms***)**

**Onset Peak**

I

1.0

57

2.8

3.1

II

2.0

73

2.8

3.2

III

3.0

78

2.8

3.4

IV

4.0

87

2.8

3.4

FIGURE 5–39 Active and reference

interelectrode distance. Median nerve

sensory nerve action potential and

the effect of varying the interelectrode

separation. (I–IV show a sequential

increase in the electrode separation.)

COMPOUND MOTOR ACTION POTENTIAL (CMAP) (FIGURE 5–40)

•

Motor nerve studies are known as CMAPs. They are also called M waves. They represent the

­ conduction of an impulse along motor nerve fibers of a motor unit.

•

It is recorded as an evoked motor potential from a motor point within the muscle. It corre-

sponds to the integrity of the motor unit but cannot distinguish between pre- and postgangli-

onic lesions because the cell body is located in the spinal cord.NERVE CONDUCTION STUDIES

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Onset

latency

FIGURE 5–40

Compound motor

action potential.

•

•

It can be abnormal with normal

SNAPs if the lesion is proximal

to the DRG (see previous section

on SNAPs) or affecting a purely

motor nerve (Figure 5–38).

Technical considerations:

– Phases:

–

nn The potential should be

biphasic with an initial

negative deflection. If an

initial positive ­ deflection

FIGURE 5–41 Compound motor action potential electrode placement.

(I) Over the endplate region. (II) Off the endplate region.

exists, it may be due to:

nn Inappropriate placement of the active electrode from the motor point (Figure 5–41).

nn Volume conduction from other muscles or nerves

nn Anomalous innervations

– Recording electrode:

–

nn A falsely decreased amplitude and inaccurate latency can be due to:

nn The active and reference electrodes should not be too close together. If this occurs,

­ similar waveforms are recorded at both sites and rejected, decreasing the amplitude of

the ­ waveform (Figure 5–42).

nn Submaximal stimulation

nn Stimulating over thickened skin from callous formation or edema

FIGURE 5–42 Compound motor action potential

electrode placement. (I) Over the endplate region.

(II) Off the endplate region.NCS SNAP CMAP

Anatomy examined Sensory nerve fibers Motor nerve fibers, NMJ, muscle fibers

Pertinent latencies Peak or onset latency Onset latency

Amplitude measurements Peak to peak (μV) Baseline to peak (mV)

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MEASUREMENT DIFFERENCES (TABLE 5–7)

TABLE 5–7 Measurement Differences

CMAP, compound motor action potential; NCS, nerve conduction study; NMJ, neuromuscular junction; SNAP,

sensory nerve action potential.

CONDUCTION VALUES (TABLE 5–8)

•

The following normal values are proposed for the basic motor and sensory NCS. Each EDX lab

may have different normal values. Table 5–8 contains proposed normal values for basic motor

and sensory NCS.

TABLE 5–8 Normal Values Proposed for Basic Motor and Sensory Nerve Conduction Study

NERVES VALUES NERVES VALUES

Median Motor Sup. Radial Sensory

Distal latency 3.7 ± 0.3 (8 cm) Distal latency 2.3 ± 0.4 (10 cm)

Amplitude 13.2 ± 5.0 Amplitude 31.0 ± 20.0

CV 56.7 ± 0.2 CV 58 ± 6.0

Median Sensory Peroneal Motor

Distal latency 3.2 ± 0.5 (14 cm) Distal latency 4.5 ± 0.8 (8 cm)

Amplitude 41.2 ± 25.0 Amplitude 4.4 ± 1.4

CV 56.9 ± 4.0 CV below fibula head 51.6 ± 4.1

Ulnar Motor CV above fibula head 53.9 ± 4.3

Distal latency 3.2 ± 0.5 (8 cm) Tibial Motor

Amplitude 6.0 ± 1.9 Distal latency 3.4 ± 0.5 (10 cm)

CV below elbow 61.8 ± 5.0 Amplitude 11.8 ± 4.5

CV above elbow 62.7 ± 5.5 CV 53.9 ± 4.3

Ulnar Sensory Sural Sensory

Distal latency 3.2 ± 2.5 (14 cm) Distal latency 3.5 ± 0.2 (14 cm)

Amplitude 34.0 ± 12.1 Amplitude 16.6 ± 7.5

CV 57.0 ± 5.0 CV 39.6 ± 2.3

Radial Motor Sup. Per. Sensory

Distal latency 2.4 ± 0.5 (10 cm) Distal latency 2.9 ± 0.3 (14 cm)

Amplitude 14.0 ± 8.8 Amplitude 20.5 ± 6.1

CV 61.9 ± 5.9 CV 65.7 ± 3.7

CV, conduction velocity.•

•

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**H - Reflex**

m H

H-REFLEX (FIGURE 5–43)

This NCS late response is an electri-

cally evoked analogue to a monosyn-

aptic reflex. In the lower extremity, it

is the EDX equivalent of the Achilles

reflex.

It is initiated with a submaximal stimu-

lus at a long duration (1.0 msec). This

preferentially activates the IA afferent

nerve fibers, causing an orthodromic

sensory response to the spinal cord,

and then an orthodromic motor response

back to the recording electrode.

The waveform can be potentiated

with agonist muscle contraction and

abolished with antagonist contraction

FIGURE 5–43 H-reflex. The H response is obtained by stimulation

of the afferent sensory fiber (top) resulting in orthodromic

conduction to the spinal cord. In the spinal cord, there is

synaptic stimulation of the alpha motor neuron, resulting in the

evoked H response in the muscle. A rudimentary M response is

produced when a few motor axons are directly stimulated.

or increased stimulation causing colli-

sion blocking.

The morphology and latency remain constant with each stimulation at the appropriate inten-

sity. Note that a mean of 10 F-waves can be used as a surrogate for one H-reflex.

Function:

– – It reflects the response of a proximally traveling evoked potential. It is typically used to

monitor for an S1 radiculopathy in the lower extremity or a C7 radiculopathy in the upper

extremity.

Formula:

– – H-reflex = 9.14 + 0.46 (Leg length in centimeter from the medial malleolus to the popliteal

fossa) + 0.1 (age).

Latency:

– – – – – – – – – – – – Normal: 28 to 30 msec

Side to side difference: >0.5 to 1.0 msec is significant

>60 years: Adds 1.8 msec

Location:

Soleus muscle: Tibial nerve: S1 pathway

Flexor carpi radialis (FCR): Median nerve: C7 pathway

Alterations:

This waveform can be seen in all nerves of adults with an upper motor neuron (UMN;

­ corticospinal tract) lesion as well as in normal infants. It is possible to potentiate a waveform

by agonist muscle contraction, and inhibit the H-reflex by antagonist contraction.

Limitations:

– – This evaluates a long neural pathway, which can dilute focal lesions and hinder specificity of

injury location. It can be normal with incomplete lesions.

– – It also cannot distinguish between acute and chronic lesions. Once it is abnormal, it is always

abnormal.

– – Pitfall: While an absent H-reflex can be seen in an S1 radiculopathy, it is NOT a specific

finding to diagnose it. Absent H-reflexes can be seen in multiple other conditions, including

generalized peripheral neuropathies, plexopathies, and upper motor neuron lesions. It can

also be a normal finding in elderly adults.

F-WAVE (FIGURE 5–44)

The F-wave is a small late motor response occurring after the CMAP. It represents a late

response from approximately 1% to 5% of the CMAP amplitude. It is produced using a

short duration, ­ supramaximal stimulation, which initiates an antidromic motor response to the

anterior horn cells in the spinal cord, which in turn produce an orthodromic motor response

to the recording electrode.360 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

**F - Wave**

m F

FIGURE 5–44 F-wave response:

Stimulation (dot) is followed by the

source of depolarization (arrows). Initially

depolarization travels in both directions, first

directly to the muscle fiber producing the

M response, and retrograde up to the axon

and to the neuron, where it is repropagated

in a small percentage of neurons back

down the axons to produce the delayed F

response.

•

•

•

•

•

•

•

The F-wave is a pure motor response and does not

represent a true reflex because there is no synapse along

the nerve pathway being stimulated. The configuration

and latency change with each stimulation due to activa-

tion of different groups of anterior horn cells with each

­ stimulation (Figure 5–45).

Function:

– May be helpful in polyneuropathies and plexopathies

–

but not overly useful in radiculopathies

Latency:

– – – – Normal: Upper limb: 28 msec; lower limb: 56 msec

Side-to-side difference: 2.0 msec difference in the

upper limbs is significant; 4.0 msec difference in

lower limbs is significant

– – Decreased persistence (occurrence) on repetitive

stimulations correlates with a potential abnormality

Location:

– – – – It can be obtained from any muscle.

Limitations:

This evaluates a long neural path-

way, which can dilute focal lesions

and hinder specificity of injury

location.

– – It only accesses the motor fibers.

A-(AXON) WAVE

When performing a CMAP study, a

response can be evoked by a submax-

imal stimulation and ­ abolished with

a supramaximal level. The stimulus

can travel antidromically along the

motor nerve and becomes diverted

along a neural branch formed by

collateral sprouting due to a ­ previous

­ denervation and reinnervation

process. It typically occurs between

the CMAP and F-wave at a ­ constant

latency (Figure 5–46).

Function:

– – This waveform represents col-

lateral sprouting following nerve

damage.

FIGURE 5–45 Renshaw cell activation.

Inhibitory neurons, Renshaw cells (R)

are activated by a stimulus and, in turn,

suppress (–) firing of the alpha motor

neuron.

**Tibial Nerve**

1 M 2 F 3

**Submaximal**

**stimulation (SI)**

1 M 2 F 3

**Supramaximal**

**stimulation (SII)**

Supramax

stim S(I)

Submax

stim S(I)

**A-Wave generated Blocking occurs**

FIGURE 5–46 A-wave. (A) Arrows 1, 2, and 3 represent the A-waves.

M is the compound motor action potential and F is the F-wave. S

(I)—weak stimulus, S (II) strong stimulus. [Note: A-waves seen in S

(I) are abolished]. (B) S (I) A-wave generated, S (II) blocking occurs.NERVE CONDUCTION STUDIES

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BLINK REFLEX

(FIGURE 5–47 AND 5–48)

•

This NCS is an electrically evoked

analogue to the corneal reflex. It is

initiated by stimulating the supraor-

bital branch of the trigeminal nerve.

The response propagates into the

pons and branches to the lateral

medulla. It then branches to inner-

vate the ipsilateral and contralateral

orbicularis oculi via the facial nerve.

•

Two responses are evaluated, an

­ ipsilateral R1 and bilateral R2. The

blink is associated with the R2

response (Table 5–9).

•

Latency (Figure 5–49A and B):

– – – – Normal: R1 <13 msec

R2 Ipsilateral (direct) <40 msec

– – R2 Contralateral (consensual)

<41 msec

FIGURE 5–47 Blink reflex procedure. Both orbicularis oculi muscles

are recorded simultaneously. The active recording electrodes (G1)

are placed inferior and slightly lateral to the pupil at mid-position,

with the reference recording electrodes (G2) placed just lateral to

the lateral canthus. For each side, the ipsilateral supraorbital nerve

is recorded over the medial eyebrow. Recording and stimulation

sites are shown for a right-sided blink reflex.

V1

VM

VII VII

VII

VII

VS

FIGURE 5–48 Blink reflex anatomy. The afferent loop of the

blink reflex is mediated by the first division of the trigeminal

nerve (V1), which synapses with both the main sensory

nucleus of cranial nerve V (VM) in the midpons and the

nucleus of the spinal tract of cranial nerve V (VS) in the

medulla. The earlier R1 potential is mediated by a disynaptic

connection between the main sensory nucleus and the

ipsilateral facial motor nucleus (VII). The later R2 responses

are mediated by a multisynaptic pathway between the

nucleus of the spinal tract of cranial nerve V and both

ipsilateral and contralateral facial nuclei (VII). The efferent

pathway for both R1 and R2 is mediated via the facial nerve

to the orbicularis oculi muscles.

TABLE 5–9 Blink Reflex Pathways

PATHWAY NERVE FIBERS

Afferent Sensory branches of CN V (trigeminal nerve)

Efferent Motor branches of CN VII (facial nerve)

CN, cranial nerve.

RESPONSE COURSE

R1 (Early) Through the pons

R2 (Late) Through the pons and lateral medulla

R1 IS AFFECTED BY LESIONS OF THE: R2 IS AFFECTED BY:

Trigeminal nerve

Pons

Facial nerve

Consciousness level

Parkinson’s disease

Lateral medullary syndrome

Valium

Habituation362 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

FIGURE 5–49A (A) Blink reflex responses. The pons

(1) is the pathway for R1. The pons and lateral

medulla (2 and 3) is the pathway for R2.

**A**

**B**

**C**

**D**

**E**

**F**

**G**

**H**

R

L

R

L

R

L

R

L

R

L

R

L

R

L

R

L

V1 Stimulation site

Right Left

R

L

R

L

R

L

R

L

R

L

R

L

R

L

R

L

FIGURE 5–49B Blink reflex patterns of

abnormalities.

(A) Normal pattern. Recording both

orbicularis oculi muscles; stimulating the

supraorbital nerve on each side results in an

ipsilateral R1 (early) and bilateral R2 (late)

potential.

(B) Incomplete right trigeminal lesion.

Stimulating the affected right side, there is a

delay of all potentials, including the ipsilateral

R1 and R2 and contralateral R2. Stimulating

the unaffected side results in all normal

potentials.

(C) Complete right trigeminal lesion.

Stimulating the affected right side, all

potentials are absent. Stimulating the

unaffected side results in all normal potentials.

(D) Incomplete right facial lesion.

Stimulating the affected side results in delay

of the ipsilateral R1 and R2, but a normal

contralateral R2. Stimulating the unaffected

side results in a normal ipsilateral R1 and

R2, but a delayed contralateral R2. In this

pattern, all potentials on the affected side

are abnormal, regardless of which side is

stimulated.

(E) Complete right facial lesion. Stimulating

the affected side results in absent ipsilateral R1

and R2 potentials, but a normal contralateral

R2. Stimulating the unaffected side results in

a normal ipsilateral R1 and R2, but an absent

contralateral R2.

(F) Right midpontine lesion (main sensory

nucleus V and/or lesion of the pontine

interneurons to the ipsilateral facial nerve

nucleus). Stimulating the affected side results

in an absent or delayed R1, but an intact

ipsilateral and contralateral R2. Stimulating the

unaffected side results in all normal potentials.

(G) Right medullary lesion (spinal tract

and nucleus V, and/or lesion of the medullary

interneurons to the ipsilateral facial nerve

nucleus). Stimulating the affected side

results in a normal R1 and R2, but an absent

or delayed ipsilateral R2. Stimulating the

unaffected side results in normal ipsilateral

R1 and R2 potentials, but a delayed or absent

contralateral R2.

(H) Demyelinating peripheral

polyneuropathy. All potentials of the blink

response may be markedly delayed or absent,

reflecting slowing of either or both motor and

sensory pathways.

DIRECT FACIAL NERVE STUDY (FIGURE 5–50)

•

NCS of cranial nerve (CN) VII (facial nerve) is performed by stimulating it distal to the stylo-

mastoid foramen at the angle of the mandible. The response is recorded over the nasalis muscle.

•

The patient can present with equal weakness in the upper and lower facial muscles with a

peripheral nerve injury.

•

If the lesion is rostral to the facial nerve nucleus (central), the lower facial muscles are more

severely affected than the upper.NERVE CONDUCTION STUDIES

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Synkinesis

An aberrant regeneration of axons can occur with facial nerve injuries leading to ­ reinnervation of

inappropriate muscles. This may present as lip twitching when ­ closing an eye or crocodile tears

when chewing.

NCS AMPLITUDE/PROGNOSIS

CMAP <10% of the unaffected side

Poor outcome, likely to incomplete recovery. Recovery >1 year

CMAP Between 10% and 30% of the unaffected side

Fair prognosis. Recovery within 2–8 months

CMAP >30% of the unaffected side

Good prognosis. Recovery within 2 months

•

Function:

– This monitors for injury to the facial nerve such as seen

–

in Bell’s palsy, neoplasms, fractures, middle ear infection,

diabetes mellitus, mumps, Lyme disease, and so on.

•

Findings:

– This can be monitored periodically over 2 weeks to

–

assess prognosis. Better outcomes are ­ anticipated for

demyelinating versus axonal injuries. Absence of an

evoked potential in 7 days indicates poor prognosis.

Treatment:

•

Interventions may include prednisone, massage, or

electrical stimulation.

Reference

electrode

Active

electrode

Ground

Anode Cathode

FIGURE 5–50 Facial nerve study.

Direct Facial Nerve

CMAP, compound muscle action potential; NCS, nerve conduction study.

PHRENIC MOTOR STUDY

•

NCS of phrenic nerve is performed by stimulating posterior to the sternocleidomastoid muscle,

approximately 3 cm above the clavicle, recording the diaphragm with G1 placed two ­ fingerbreadths

above the xiphoid process and G2 placed over the anterior costal margin 16 cm from G1.

•

•

The diaphragm CMAP is usually biphasic, with the initial phase upward.

Needle EMG of the diaphragm:

– – The diaphragm can be studied with a needle EMG electrode, using three approaches: sub-

costal, lower lateral intercostals, and substernal. The subcostal approach is the most com-

monly used for the reasons of safety and assurance of proper needle position.

– – In the subcostal approach, the EMG needle electrode is inserted under the costal margin

behind the eighth, ninth, or tenth rib cartilage.

– – It is important to confirm the location of the EMG needle in the diaphragm by observing

­ inspiratory electrical activity.

– – The presence of fibrillation potential (FIB) and PSWs is strongly suggestive of neurogenic

impairment. In a partially denervated diaphragm, FIBs and PSWs are observed between

the bursts of inspiratory electrical activities. In a complete denervated diaphragm, there is

absence of inspiratory electrical activity, and only FIBs and PSWs may be seen.

•

Treatment for diaphragm paralysis:

– – Phrenic pacing is increasingly being used in patients with central respiratory paralysis and

upper cervical spinal cord injury (lesions above C3) to wean them off the ventilators. These

patients ­ ideally should not have any intrinsic lung disease.

– Electrodes can be implanted intrathoracically via thoracotomy and, more recently, with

–

video-assisted thoracoscopic surgery (VATS). Alternatively, electrodes can be placed intra-

muscularly via a laparoscopic approach. In this approach, intramuscular electrodes are

placed near the entrance points of the phrenic nerves using motor-point mapping techniques.364 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

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SOMATOSENSORY EVOKED POTENTIALS (FIGURE 5–51)

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This study evaluates time-locked responses of the nervous system to an external stimulus. They

represent the function of the ascending sensory pathways using an afferent potential, which

travels from the peripheral nerve to the plexus, root, spinal cord (posterior column), contralat-

eral medial lemniscus, thalamus, to the somatosensory cortex.

It is initiated by a repetitive submaximal stimulation of a sensory nerve, mixed nerve, or

dermatome, and is recorded from the spine or scalp. The nerves most commonly used for

testing are the median for the upper limb and the tibial nerve for the lower limb.

Function:

– Somatosensory evoked potential (SSEP) monitors for problems such as (a) peripheral

–

nerve injuries, (b) CNS lesions such as multiple sclerosis (MS), or (c) intraoperative monitor-

ing of spinal surgery.

– – Changes in MS are seen 90% of the time, with the lower limb more likely to be abnormal

than the upper limb. The most common abnormality is the prolonged interpeak latencies.

Amplitude reduction or absence can also be seen.

– During spinal cord surgery, loss of tibial nerve potentials with preservation of median

–

nerve potentials can indicate nerve injury at the level of intervention. Anesthesia will affect

SSEP ­ potentials in both the upper and lower limbs.

Paracentral lobule or

Laterally depending if upper

or lower extremity is stimulated

Projection fibers to

parietal lobe

VPL Nucleus

Medial lemniscus

Nucleus gracilis or

nucleus cuneatus

Posterior columns

Internal arcuate fibers

Peripheral nerve

–

+

Stimulating electrodes

FIGURE 5–51 Somatosensory-evoked potential pathways from the peripheral nerve to the parietal cortex. The

fasciculus gracilis is monitored when a lower extremity nerve is stimulated and the fasciculus cuneatus is monitored

when an upper extremity nerve is stimulated.SOMATOSENSORY EVOKED POTENTIALS

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Recording Sites

Median nerve SSEP (Figure 5–52):

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N9—Erb’s point (reflects brachial plexus integrity)

•

N11—Roots

•

N13—Cervicomedullary junction (nucleus cuneatus)

•

P14—Lower brainstem

•

N18—Rostral brainstem

•

N20—Primary cortical somatosensory receiving area

Tibial nerve SSEP (Figure 5–53):

•

PF—Popliteal fossa

•

L3—Third lumbar

•

N22—T12 and lumbosacral spine

•

N45—Cortical

SSEP, somatosensory evoked potential.

STIM

N20

F2

C3‘

C5S

EP1

EP2

N20

P9

P11

P13-14

N11 N13

N9

N9

–

0.2 µV

+

–

0.5 µV

+

+ –

STIM

0 10 20 30 40

50 msec

FIGURE 5–52 Median nerve somatosensory evoked potential.

N45

Fpz‘

Cz

‘

STIM

P37

T12S

L3S

–

2 µV

+

0 10 20 30 40 50 msec

+ –

STIM

FIGURE 5–53 Tibial nerve somatosensory evoked potential.366 •

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5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

Advantages:

– – SSEP theoretically evaluates the sensory components of the peripheral nervous system (PNS)

and CNS. It can aid in studying disorders of the CNS (brain, brainstem, spinal cord) as well

as of ­ dorsal nerve roots and peripheral nerves when severe peripheral disease is noted.

Abnormal results present immediately.

Limitations:

– – It only evaluates the nerve fibers sensing vibration and proprioception.

– – It is also limited in its ability to localize a nerve lesion to a focal area. It evaluates a long neu-

ral pathway, which may dilute focal lesions and hinder specificity of injury location.

– – It can be adversely affected by sleep and high doses of general anesthetics (halothane, enflu-

rane, isoflurane). This may be avoided with nitrous oxide or low dose isoflurane.

•

n

BASIC NEEDLE EMG

Needle EMG assesses nerve and muscle function. A recording needle electrode is placed into a

muscle to evaluate the following parameters:

– – Insertional activity

– – Resting activity

– – Voluntary recruitment

•

•

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•

INSERTIONAL ACTIVITY (FIGURE 5–54 AND TABLE 5–10)

Insertional activity represents discharge potentials that are mechanically provoked by

­ physically ­ disrupting the muscle cell membrane with a needle electrode. This is an electrical

injury potential.

Increased insertional activity may be seen in both neuropathic and myopathic conditions.

In rare conditions, where significant muscle atrophy has occurred, insertional activity may be

decreased.

Severe, acute ischemia of muscle due to vascular occlusion or compartment syndrome may also

produce decreased or absent insertional activity.

**A**

**B**

FIGURE 5–54 Insertional activity. (A) Normal. (B) Increased.

TABLE 5–10 Insertional Activity

CHARACTERISTIC NORMAL INCREASED DECREASED

Duration 300 msec >300–500 msec <300 msec

Etiology •

Muscle depolarization • Denervation

•

Irritable cell membrane

•

•

•

•

Fat

Fibrosis

Edema

Electrolyte

abnormalitiesBASIC NEEDLE EMG

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RESTING ACTIVITY

Normal Spontaneous Activity (Figure 5–55 and Table 5–11)

•

After a needle is inserted into a normal muscle, there should be electrical silence at rest.

However, if it is placed into or near the NMJ (motor endplate), two waveforms can occur:

MEPPs and EPPs. Needle placement in this area is painful, but these waveforms also indicate

that the muscle has ­ maintained its innervation.

– MEPPs occur spontaneously at the NMJ, and are referred to as endplate noise.

–

nn They result from the normal spontaneous exocytosis of individual quanta of ACh travel-

ing across the NMJ, leading to a nonpropagated, subthreshold EPP. They have a distinc-

tive small amplitude of 10 to 50 μV and monophasic negative morphology.

nn This “endplate noise” is due to spontaneous quanta release (100–200 quanta), which occurs

every 5 seconds. It results in a 10 to 50 μV, nonpropagated potential seen on the screen as

an irregular baseline when it is recorded with a standard extracellular needle electrode.

– EPPs are endplate spikes due to increased ACh release, provoked by needle irritation of the

–

­ muscle fiber or synchronization of several MEPPs. It results in a propagated single muscle

fiber AP.

nn Its hallmark sign is its irregularity and it always presents as a negative deflection. It can

be misinterpreted as a positive wave if the needle is moved a little bit.

A B

CHARACTERISTICS MEPP EPP

Initial deflection Negative (monophasic) Negative (biphasic)

Duration 0.5–1.0 msec 2.0–4.0 msec

Amplitude 10–50 µ V <1,000 µ V (1 mV)

Rate 150 Hz 50–100 Hz

Rhythm Irregular Irregular

Origin Endplate Endplate/provoked

Sound Sea shell murmur Sputtering fat in a frying pan

FIGURE 5–55 Endplate activity: (A) MEPP—Monophasic negative potentials primarily present as baseline

irregularities because of their small amplitude. (B) EPP—Biphasic, usually negative single-fiber action potential.

EPP, endplate potential; MEPP, miniature endplate potential.

TABLE 5–11 Normal Spontaneous Activity

EPP, endplate potential; MEPP, miniature endplate potential.

Abnormal Spontaneous Activity

•

Pathologic waveforms can be generated from a muscle fiber or from the motor unit (neu-

ral source). They can be caused by a variety of conditions, including muscle denervation or

myopathies (­ inflammation, dystrophies). If originating from a muscle source, the activity can

represent lack of muscle fiber innervation.368 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

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The nerve RMP becomes less negative and

unstable, causing it to approach the thresh-

old more easily to activate an AP. It either

fires independent of external stimulation or

is induced by needle ­ movement. If neural in

origin, it can have the appearance of MUAPs

(Figure 5–56).

Abnormal spontaneous activity from muscle

fibers includes:

– – FIBs

– – PSWs

– – Complex repetitive discharges (CRDs)

– – Myotonic discharges

Abnormal spontaneous activity from a motor

unit includes:

– – Fasciculation potentials

– – Myokymic discharges

– – Neuromyotonic discharges

1. ABNORMAL SPONTANEOUS ACTIVITY

GENERATED FROM MUSCLE FIBER OR NERVE

PATHOLOGY (TABLES 5–12 AND 5–13)

•

Fibrillations (FIBs) (Figure 5–57):

FIGURE 5–56 Spontaneous waveform source

generators. Spontaneous activity originates from

a variety of source generators. Each generator is

associated with a specific morphology.

– – Spontaneously firing APs originating from

denervated single muscle fibers secondary to ­ uncontrolled ACh release.

– – Although they are typically associated with neuropathies, they can also be seen in

myopathies.

– – On needle EMG, hallmarks are sounding like “rain on a tin roof” and their regularity of

firing.

•

Positive sharp waves (PSWs) (Figure 5–58):

– – Similar to FIBs, PSWs represent spontaneous firing of single muscle fibers.

– It is a needle recording of an AP of a single muscle fiber. There is propagation to but not

–

past the needle tip. This inhibits the display of the negative deflection of the waveform.

– – Same pathological significance as FIBs.

– – Hallmarks on EMG are sounding like dull thuds or pops and is also their regularity of firing.

– – Similar to FBIs, they can be found in ­ neuropathies as well as myopathies.

Table 5–12 Characteristics of Fibrillations and Positive Sharp Waves

FIBs, fibrillation potentials; PSWs, positive sharp waves.

CHARACTERISTICS FIBs PSWs

Initial Deflection and Morphology Positive (biphasic) Positive (biphasic)

Duration 1–5 msec 10–30 msec

Amplitude Early: >300 µ V Late: <25 µV <1 mV

Rate 1–10 Hz 1–20 Hz

Rhythm Regular Regular

Origin Postjunctional Postjunctional

Sound Rain on a tin roof Dull thud or chugBASIC NEEDLE EMG

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TABLE 5–13 Distribution and Intensity of Fibrillation Potentials and Positive Sharp Waves

GRADE CHARACTERISTICS

0 None

1+ Persistent single runs >1 second in two areas

2+ Moderate runs >1 second in three or more areas

3+ Many discharges in most muscle regions

4+ Continuous discharges in all areas of the muscle

FIGURE 5–57 Fibrillation potentials. FIGURE 5–58 Positive sharp waves.

•

•

ETIOLOGY:

nn Nerve disorders: Anterior horn cell disease, radiculopathy, plexopathy, peripheral neuropa-

thy, mononeuropathy

nn NMJ disorders: Myasthenia gravis, botulism

nn Muscle disorders: Muscular dystrophies, polymyositis, dermatomyositis, hyperkale-

mic p­ eriodic paralysis, acid maltase

deficiency

Complex repetitive discharges (CRDs)

(Figure 5–59 and Table 5–14):

– Formerly known as“bizarre high

–

­ frequency discharges,” CRDs are sponta-

neous, polyphasic/­ serrated APs originat-

ing from a principal pacemaker, initiating

a group of single muscle fibers to fire in

near synchrony.

The current spreads to the other muscle fibers

by ephaptic transmission (Figure 5–60). This

results from a process in which denervated

muscle fibers are reinnervated by collateral

FIGURE 5–59 Complex repetitive discharges.370 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

TABLE 5–14 Complex Repetitive Discharges

FIBs, fibrillation potentials; MUAP, motor unit action potentials; PSWs, positive sharp waves.

CHARACTERISTICS COMPLEX REPETITIVE DISCHARGES

Morphology Resembles FIBs, PSWs, MUAP

Amplitude 50–1,000 µ V

Rate 10–100 Hz

Rhythm Regular/abrupt start and stop

Origin Postjunctional/ephaptic transmission

Sound Motor boat

B

A

C

D

2

1

3

4

FIGURE 5–60 Ephaptic transmission. Fiber A acts as the pacemaker. It transmits an impulse to B, C, and D, which

perpetuates the cycle.

sprouting from axons of a n­ eighboring motor unit. When these fibers, in turn, become dener-

vated, a population of muscle fibers now belonging to one motor unit lacks neural control.

These muscle fibers lie in close ­ proximity to each other and serve as a circuit for the pace-

maker fiber.

– Its hallmark sign is its regular interval between each discharge and within each discharge and

–

a “motor boat engine” sound.

ETIOLOGY

nn Nerve disorders: Anterior horn cell ­ disease, chronic radiculopathy, peripheral neuropathy

nn Muscle disorders: Polymyositis, ­ dermatomyositis, muscular dystrophies, limb-girdle dys-

trophy, myxedema

nn Normal variant

•

Myotonic discharges (Figure 5–61 and

Table 5–15).

– – These are biphasic single muscle fiber

APs triggered by needle movement,

percussion, or voluntary contraction.

FIGURE 5–61 Myotonic discharges.

They are caused by an alteration of the

ion channels in the muscle membrane and can be seen with or without clinical myotonia. Its

hallmark sign is the smooth change in rate and amplitude with a “dive bomber” sound.

ETIOLOGY

nn Nerve disorders: Chronic radiculopathy, peripheral neuropathy

nn Muscle disorders: Myotonic dystrophy, myotonia congenita, paramyotonia, polymyositis,

­ dermatomyositis, acid-maltase deficiency, hyperkalemic periodic paralysis

nn Medications: PropranololCHARACTERISTICS MYOTONIC DISCHARGES

Morphology Resembles: Single-muscle-fiber action potential

Duration >5–20 msec

Amplitude 20–300 µ V

Rate 20–100 Hz

Rhythm Wax and wane

Origin Postjunctional

Sound Dive bomber

CHARACTERISTICS FASCICULATIONS

Morphology Resembles MUAP with long, irregular intervals

Duration 5–15 msec

Amplitude <300 µ V

Rate <1–2 Hz

Rhythm Irregular

Origin Prejunctional

Sound Corn popping—dull, irregular pops

BASIC NEEDLE EMG

TABLE 5–15 Myotonic Discharges

2. ABNORMAL SPONTANEOUS ACTIVITY GENERATED

FROM THE MOTOR UNIT (NEURAL SOURCE)

•

Fasciculations (Figure 5–62 and Tables 5–16 and 5–17):

– – These are spontaneous discharges originating from the

motor neuron or its axon prior to its terminal branches,

and result in intermittent muscle fiber contraction.

If associated with FIBs or PSWs, they are considered

pathological.

– A fasciculation has the appearance of a MUAP

–

with an irregular firing pattern. Its hallmark sign

is a much slower frequency with a “corn popping”

sound. Fasciculations can be pathologic or benign.

Differentiating between the two on testing alone is

nearly impossible. However, benign fasciculations

are not associated with muscle weakness, atrophy, or

reflex abnormalities.

FIGURE 5–62 Fasciculations.

ETIOLOGY

nn Nerve disorders: Anterior horn cell disease, tetany, Creutzfeldt–Jakob syndrome, radicu-

lopathy, mononeuropathy

nn Metabolic disorders: Thyrotoxicosis, tetany

nn Normal variant

TABLE 5–16 Fasciculations

MUAP, motor unit action potential.

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TABLE 5–17 Distribution and Intensity of Fasciculations

GRADE CHARACTERISTIC

0 None

+/– Equivocal

1+ In two areas, 2–10 per minute

2+ In many areas, 10–15 per minute

3+ All areas, <60 per minute

4+ All areas, >60 per minute

•

Myokymic discharges (Figure 5–63 and Table 5–18):

– – These are rhythmic groups of MUAPs firing repetitively. They can be associated with clinical

myokymia, which presents as slow continuous muscle fiber contractions. This gives a rip-

pling appearance to the overlying skin.

– – Myokymic discharges are seen in multiple conditions but are most commonly seen in

radiated-induced neuropathy.

– – Its hallmark sign is the semiregularity between each discharge and within each discharge

with the characteristic sound of “marching soldiers.”

ETIOLOGY

nn nn Facial myokymia: MS, brainstem neoplasm, polyradiculopathy, Bell’s palsy

Extremity myokymia: Radiation plexopathy (most common), compression neuropathy, rattle-

snake venom

FIGURE 5–63 Myokymic discharges.

TABLE 5–18 Myokymic Discharges

CHARACTERISTICS MYOKYMIC DISCHARGES

Morphology Bursts of MUAPs

Amplitude 100 µ V–2 mV

Rate Discharge: 40–60 Hz; Interdischarge: 0.1–10 Hz

Rhythm Semiregular

Origin Nerve

Sound Marching soldiers

MUAP, motor unit action potential.BASIC NEEDLE EMG

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•

Neuromyotonic discharges (Figure 5–64 and Table 5–19):

– – These are high-frequency, repeti-

tive discharges with progres-

sively decremental amplitudes

­ originating from damaged

peripheral motor axons. The

progressive decrement of their

­ waveforms is due to individual

muscle fiber fatigue and drop

off.

– – They are classically seen in neu-

romyotonia (Isaac’s syndrome).

This is a disorder associated

with continuous muscle fiber

activity resulting in the appear-

ance of muscle rippling and

stiffness ­ secondary to irritable

nerves.

– – Hallmark signs on EMG include

high-frequency, repetitive

discharges with progressively

­ decremental amplitudes with a

characteristic “pinging” sound

on EMG.

ETIOLOGY

nn Nerve disorder: Neuromyotonia, chronic neuropathic disease, tetany

nn Toxins: Anticholinesterase

FIGURE 5–64 Neuromyotonic discharges in spinal muscular atrophy.

TABLE 5–19 Neuromyotonic Discharges

CHARACTERISTICS NEUROMYOTONIC DISCHARGES

Morphology High-frequency, repetitive discharges with progressively decremental amplitudes

Amplitude Progressively decremental

Rate 150–250 Hz

Rhythm Regular; discharges continuously or in bursts

Sound “Pinging”

Appearance Tornado-like

3. CRAMP DISCHARGES (FIGURE 5–65 AND TABLE 5–20)

•

These are spontaneous discharges associated with repetitive firing of MUAPs in a large area of

muscle. They originate from high-frequency discharges from motor axons and are not muscle in

primary origin.

•

They are usually associated with painful, involuntary muscle contractions and are

synchronous.

ETIOLOGY

nn Electrolyte disturbances, uremia, pregnancy, myxedema, strenuous exercises, ­ prolonged

muscle contraction, liver cirrhosis, myotonia congenita, myotonic dystrophy, stiff-man’s

syndrome.374 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

TABLE 5–20 Cramp Discharges

MUAP, motor unit action potential.

CHARACTERISTICS CRAMP DISCHARGES

Morphology Multiple, normal MUAPs firing at a high, irregular

frequency

Duration Abrupt onset and cessation

Amplitude Up to 1 mV

Rate 40–60 Hz

Rhythm Irregular

FIGURE 5–65 Muscle cramp discharges.

4. ARTIFACT POTENTIALS (FIGURES 5-66 AND 5-67)

•

These are waveforms that obscure the neurophysiologic signals. Interference potentials are

unwanted signals occurring from outside the system being studied.

•

Noise is described as unwanted signals occurring from within the system. They arise from the

EMG instrument, printer, unshielded power cords, electrical outlets, fluorescent lights, or a

pacemaker.

20 µV

5 ms

FIGURE 5–66 Source of noise and interference.

FIGURE 5–67 Fluorescent light interference.

Noise is greatest near electrical outlets,

Source:From Dumitru D. Electrodiagnostic Medicine.

power cords, and equipment including the

Philadelphia, PA: Hanley & Belfus; 1995, with

electromyography instrument itself.

permission.

EXERTIONAL ACTIVITY

•

•

Voluntary muscle fiber activity that can be electrically recorded and summated as a MUAP.

Motor Unit Action Potential (MUAP) (Figure 5–68 and Table 5–21):

– – This is a compound action potential from muscle fibers belonging to a single motor unit,

within the recording range of the needle electrode. Its territory is typically 5 to 15 mm.BASIC NEEDLE EMG

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Turns

Amplitude

Satellite

Phase

Baseline crossing

Rise

time

Duration

FIGURE 5–68 Motor unit action potential.

CHARACTERISTICS MUAP

Initial deflection Positive/negative

Duration 5–15 msec

Amplitude 1 µ V to 2 mV

Rise time <500 msec

Phases 2–4

Rate Exertion dependent

Rhythm Exertion dependent

Origin Prejunctional

TABLE 5–21 Motor Unit Action Potentials

MUAP, motor unit action potential.

Parameters

•

Amplitude:

– This represents the muscle fibers recorded near the needle electrode. It is measured from the

–

most positive to the most negative peak. It can be increased from a reinnervation process,

decreased from loss of muscle fibers, or variable due to blocking associated with NMJ disorders.

– – Normal: 1 mV

•

Rise time:

– – This represents the time it takes the MUAP to go from its baseline to the peak of the negative

wave. It represents proximity of the needle to the motor unit.

– – Normal: <500 μsec

•

Duration:

– – This represents the number of muscle fibers within the motor unit. It is measured from the

waveform’s initial departure from baseline to its final return. It increases (>15 msec) as the

motor unit territory increases from collateral sprouting or decreases (<5 msec) with the loss

of muscle fibers.

•

– – – – Normal: 5 to 15 msec

Turns:

These are changes in the direction of the waveform that do not cross the baseline. They are

also called serrations.

•

Phases:

– – This represents the synchronicity of muscle fiber APs firing. They are calculated as the num-

ber of baseline crossings plus 1.376 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

– – – – Five or more baseline crossings represent polyphasicity. This can result from pathology caus-

ing muscle fiber dropout, alterations in fiber CV, or reinnervation from collateral sprouting.

nn However, this can occur normally 15% (concentric needle) or 30% (monopolar needle) of

the time in adults, or more frequently in the elderly.

Normal: 2 to 4

Abnormalities

•

Long-duration, large-amplitude (LDLA) polyphasic potentials:

– These potentials typically occur as a result of denervation and reinnervation from collat-

–

eral sprouting. This reinnervation process causes an increased number of muscle fibers per

motor unit.

– – It is most commonly seen in neuropathic diseases but can also occur in chronic myopathic

disease from fiber splitting, as seen in an inflammatory or dystrophic myopathy. The recruit-

ment patterns help distinguish between a myopathic or neuropathic process.

•

Short-duration, small-amplitude (SDSA) polyphasic potentials:

– – These potentials typically occur from a dropout or dysfunction of muscle fibers. It is most

­ commonly seen in myopathic diseases and NMJ disorders. It can also occur from a severe

­ neuropathic injury leading to nascent motor unit potentials resulting from axon regrowth.

•

– – – – Again, the two processes are differentiated by their recruitment patterns.

Neuropathic potentials:

This term is not considered to be accurate because certain disease processes mimic MUAP

features, regardless of whether the pathology originated in nerve or muscle. It refers to

LDLA MUAPs that occur from a denervation and reinnervation process typically seen in

neuropathies.

– – This terminology also lacks the appropriate quantitative description of the motor unit

parameters.

•

Myopathic potentials:

– – This term is also not considered accurate because certain disease processes mimic MUAP

features, regardless if the pathology is muscle or nerve in origin. It refers to SDSA MUAPs

that occur from muscle disorders.

– – This terminology lacks the appropriate quantitative description of the motor unit

parameters.

•

Unstable potentials:

– – – – This refers to the occurrence of variations in the MUAP’s amplitude, duration, and slope.

It is most commonly seen in NMJ disorders, which cause irregular blocking of discharges.

It also can occur in motor neuron disorders, neuropathic disorders, or muscle trauma and

reinnervation.

•

Satellite potentials:

– – – – These are small potentials seen in early reinnervation.

They are time-locked potentials that trail the main MUAP . It can be due to the newly formed

­ neural sprouting that often is small, unmyelinated or thinly myelinated ® very slowly

conducting.

– – These satellite potentials are extremely unstable and may vary slightly in their firing rate

(FR) or may block and not fire at all. Over time, as the sprout matures and the CV increases,

the satellite potential will become an additional phase or serration within the main complex.

•

Doublet/multiple potentials:

– – This refers to two or more MUAPs firing recurrently and together in a semirhythmic fashion.

It is seen in ischemia, hyperventilation, tetany, motor neuron disorder, or metabolic diseases.

•

Giant potentials:

– – This refers to the extremely large MUAPs (>5 mV) that occur in disease processes such as

­ poliomyelitis. These are also described as large amplitude potentials.

RECRUITMENT (TABLE 5–22)

•

Recruitment refers to the ability to add successive motor units to increase the force of a contrac-

tion. With normal minimal contraction, a single MUAP fires before a second MUAP is recruited.BASIC NEEDLE EMG

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A B C

FIGURE 5–69 Motor unit recruitment. (A) Normal. (B) Early (myopathic). (C) Decreased (neuropathic).

TABLE 5–22 Normal Motor Unit Action Potential Recruitment Pattern

FIRST (A) SECOND (B) THIRD (C) FOURTH (D)

A—5 Hz

A—10 Hz B—5 Hz

A—15 Hz B—10 Hz C—5 Hz

A—20 Hz B—15 Hz C—10 Hz D—5 Hz

Motor Unit Recruitment Patterns (Figure 5–69)

•

Normal MUAP recruitment pattern—The rule of fives:

– – The onset frequency of the first MUAP begins at approximately 5 Hz. To generate more

force, the FR and recruitment of more motor units must be increased.

– – When the FR reaches approximately 10 Hz, a second MUAP begins at approximately 5 Hz.

– – When the first MUAP reaches a rate of 15 Hz, the second should be at 10 Hz and a third will

begin at 5 Hz.

– – As more force is needed, the FR of the first may reach 20 Hz, the second 15 Hz, the third 10

Hz, and a fourth will begin at approximately 5 Hz.

•

Early recruitment (myopathic):

– – Refers to abnormally early firing of many MUAPs with a mild contraction.

– – It is most commonly seen in myopathic conditions that result in a loss of muscle fibers as

well as some NMJ disorders. This loss causes less force to be generated per motor unit. Thus,

more motor units must now be called upon.

•

Reduced recruitment (neuropathic):

– – Refers to the firing of fewer than expected MUAPs with even a maximal contraction.

– – It is most commonly seen in neuropathic conditions but can also be in severe myopathies.

Recruitment Parameters

•

Firing Rate (FR) (Figure 5–70):

– – This is the number of times a MUAP fires per second. It is expressed in Hz and is calculated

by dividing 1,000 by the interspike interval (II) measured in msec. FR = 1,000/II.

15 ms 10 ms FIGURE 5–70 Firing rate: 1,000/13 =

approximately 75.

•

Recruitment frequency (RF):

– – This is the FR of the first MUAP when a second MUAP begins to fire. It is initiated by an

increase in the force of a contraction. Normal is considered 20 Hz or below. Values above this

correlate with a neuropathic process.FIRING PATTERN NEUROPATHY MYOPATHY

Recruitment interval ¯ ­

Recruitment frequency ­ ¯

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•

Recruitment interval (RI):

– – This is the interspike interval (in milliseconds) between two discharges of the same MUAP

when a second MUAP begins to fire. It is initiated by an increase in the force of a contraction.

Normal is considered approximately 100 msec.

RI Versus RF:

– – The following parameters help distinguish between a neuropathic and myopathic process:

– Decreased RI (Increased RF):

–

nn A loss of motor units restricts additional motor unit activation to increase contractile

force. This causes the first motor unit to fire more rapidly until a second motor unit finally

joins in. This shortens the interval between successive MUAPs from one motor unit.

– Increased RI (Decreased RF):

–

nn A loss of muscle fibers causes a second motor unit to join in early to help increase ­ contractile

force. This occurs before the first motor unit has the opportunity to increase its firing

­ frequency. This lengthens the interval between successive MUAPs from one motor unit.

– Recruitment ratio (RR):

–

nn This has been used to represent

­ recruitment capabilities, especially

when a patient ­ demonstrates difficulty

in controlling a contractile force. It is

calculated by dividing the FR of the

first MUAP by the number of different

MUAPs on the screen. A motor unit

­ firing at 10 Hz when two different

MUAPs are viewed on the screen

demonstrates an RR of 5. 10 Hz/2

FIGURE 5–71 Recruitment ratio: 10 Hz/2 different

MUAP = 5.

MUAP , motor unit action potential.

­ different MUAP = 5. The normal RR is

considered <10 (Figure 5–71).

INTERFERENCE PATTERN (FIGURE 5–72)

•

•

This is a qualitative or quantitative description of the sequential appearance of MUAPs.

It is the electrical activity recorded from a muscle during a maximum voluntary contraction. It

is composed of recruitment plus activation.

•

Activation is the ability of a motor unit to fire faster to produce a greater contractile force and is

controlled by a central process.

•

•

It can be decreased in CNS diseases, pain, and hysteria.

Furthermore, if a patient is asked to generate a force and only a few MUAPs are seen while the

­ frequency (Hz) continues to remain low, it can indicate decreased activation from poor patient

­ cooperation and is not the result of abnormal recruitment.

MUAP, motor unit action potential.

PATTERNS (FIGURE 5–72) PRESENTATION

Complete No individual MUAPs can be seen. A full screen represents four to five MUAPs

Reduced Some MUAPs are identified on the screen during a full contraction

Discrete Each MUAP can be identified on the screen during a full contraction

Single unit One MUAP is identified on the screen during a full contractionRADICULOPATHY

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Rest Voluntary Contraction

Weak Strong

500 µV

0.5s

–

+

FIGURE 5–72 Interference patterns: Representation of normal recruitment patterns.

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RADICULOPATHY

GENERAL (FIGURE 5–73)

•

This is a pathologic process affecting the nerves at the root level. In descending order, it most

­ commonly presents as pure sensory complaints, sensorimotor complaints, or pure motor com-

plaints. This involves multiples, as each dermatome is innervated by one spinal nerve root due

to the larger size of the sensory fibers, rendering them more prone to injury.

•

NCSs are typically normal. A pure sensory injury would demonstrate a negative EMG. Sensory

NCS would be normal due to sparing of the DRG.

•

It can occur without structural abnormalities on diagnostic imaging studies (MRI) or physi-

ologic abnormalities on EMG.

Spinous process

Epidural space

Dorsal horn

Lamina

Subarachnoid space

Spinal nerve

Ventral horn

Dorsal root

Dorsal root

ganglion

Dorsal ramus

Ventral

ramus

Ventral root

Body of vertebra

Epidural space

FIGURE 5–73 Cervical spine cross section.

ETIOLOGY

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Common:

– Herniated nucleus pulposus (HNP): Most common; typically seen in adults <50 years of age

–

– Spinal stenosis: Typically seen in adults >50 years of age–380 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

•

Uncommon: “HI MADAM”:

H—Herpes zoster

I—Inflammatory: Tuberculosis (TB), Lyme disease, HIV, syphilis, cryptococcus, and sarcoidosis

M—Metastasis

A—Arachnoiditis: Myelogram, surgery, steroids, and anesthesia

D—Diabetes mellitus

A—Abscess

M—

Mass in the spine: Meningioma, neurofibroma, leukemia, lipoma, synovial cysts, and

hematoma

CLINICAL FINDINGS (TABLE 5–23)

TABLE 5–23 Clinical Findings of Radiculopathies

NERVE

ROOT

REDUCED/

ABSENT REFLEX WEAKNESS NUMBNESS/PARESTHESIAS

C5 Biceps brachii Elbow flexion Lateral shoulder

C6 Brachioradialis Elbow flexion Radial forearm, thumb, index finger

C7 Triceps brachii Elbow extension Middle finger

C8 None Finger flexion Ring, little finger, hypothenar eminence

T1 None Finger adduction Ulnar forearm

L1 None Hip flexion Inguinal region

L2 None Hip flexion/knee

extension

Inguinal region/proximal anterolateral thigh

L3 Patellar tendon Hip flexion/knee

extension

Inguinal region/mid-anterolateral thigh

L4 Patellar tendon Knee extension Ankle

dorsiflexion

Anterolateral thigh/anteromedial calf/medial foot

L5 Medial hamstring Ankle dorsiflexion

Hallux extension

Foot eversion

Posterolateral thigh/calf and dorsal foot

S1 Achilles tendon Plantar flexion

Foot eversion

Posterior thigh/calf and lateral toes and heel

ELECTRODIAGNOSTIC FINDINGS

•

The guidelines for evaluation of a radiculopathy should include at least one motor and one

sensory NCS, as well as needle EMG examination including a sufficient number of muscles

representing all the relevant myotomes.

•

Nerve Conduction Studies:

– – The NCS portion of EDX studies are rarely abnormal in cases of radiculopathies. The pri-

mary role of NCS is to evaluate if there are superimposed neurologic pathologies to explain

the patient’s symptoms.

– – SNAP: Normal if the lesion is located proximal to the DRG

– CMAP: Normal or reduced amplitude. The lesion is distal to the motor neuron cell body. It can

–

be normal if the injury is purely demyelinating, incomplete, or reinnervation has occurred.

•

Late responses:

– – H-reflex: Possibly abnormal in an S1 radiculopathy but not pathognomonic

– – F-waves: Not sensitive or specific for a radiculopathy. Muscles have more than one root

­ innervation, which can result in a normal latency.RADICULOPATHY

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ROOT C3/C4 C5 C6 C7 C8

Etiology C2–C3 + C3–C4 HNP C4–C5 HNP C5–C6 HNP C6–C7 HNP C7–T1 HNP

Muscles

Involved

Clinical diagnosis

No discrete myotomal

patterns Innervates the

posterior and lateral

scalp Patient may

­ complain of ­ headaches

C2 and C3 nerves

become the greater

and lesser ­ occipital

nerve, respectively

Rhomboids

Deltoid

Biceps

Supraspinatus

Infraspinatus

Brachialis

BR

Supinator

Paraspinals

Deltoid

Biceps

BR

Supraspinatus

Infraspinatus

Supinator

PT

FCR

EDC

Paraspinals

PT

FCR

EDC

Triceps

Paraspinals

Triceps

FCU

FDP

Abductor digiti

minimi

First dorsal

interossei

PQ

Abductor pollicus

brevis

Paraspinals

•

•

Needle EMG:

– Needle EMG criteria for diagnosing a radiculopathy includes abnormal findings in two or more

–

muscles innervated by the same spinal nerve root but by different peripheral nerves. Ideally, six

muscles (five peripheral muscles + paraspinals) should be evaluated (Dillingham, 2013).

– The optimal number of muscles to screen for a cervical or lumbar radiculopathy is six (five

–

peripheral muscles + paraspinal muscles). If one of the six muscles is abnormal, then further

muscles should be evaluated. A priority is placed on including weak muscles in the evaluation.

– Classically, FIBs or PSWs should be found in two different muscles innervated by two

–

­ different peripheral nerves originating from the same root. They may not be found if the lesion

is ­ demyelinating neuropathies, pure sensory nerve injuries, chronic nerve injuries, or missed by

random sampling.

SSEP:

– Advantage: It monitors sensory pathways and proximal demyelinating injuries.

–

– Disadvantage: The long pathway monitored can mask focal lesions between the recording

–

sites, and are not considered useful for radiculopathies.

CERVICAL MYOTOMES (TABLE 5–24)

TABLE 5–24 Cervical Myotomes Effected Secondary to HNP

BR, brachioradialis; EDC, extensor digitorum communis; FCR, flexor carpi radialis; FCU, flexor carpi ulnaris; FDP,

flexor digitorum profundus; HNP, herniated nucleus pulposus; PQ, pronator quadratus; PT, pronator teres.

LUMBOSACRAL MYOTOMES (TABLE 5–25)

TABLE 5–25 Lumbar Myotomes Effected Secondary to HNP

ROOTS L2/3/4 L5 S1 S2/3/4

Etiology L1–L2/L2–L3/L3–L4

HNP

Posterolateral

L4–L5 HNP

Posterolateral

L5–S1 HNP

Iatrogenic, Cauda equina, spinal

stenosis

Muscles

involved

Iliopsoas

Iliacus

Gracilis

Adductor longus

Vastus medialis

TA

Paraspinals

Difficult to

­ distinguish between

­ radiculopathy and

alternate lesions

due to only two

­ peripheral nerves

Gluteus

maximus

Gluteus medius

TFL

TA

Medial

gastrocnemius

Medial

hamstring

TP

Palmaris longus

Paraspinals

Gluteus

maximus

Gluteus medius

TFL

Medial

gastrocnemius

Medial

hamstring

PL

TP

Paraspinals

Abductor hallucis

Abductor digiti quinti

Needle exam of the external anal

sphincter

Other clinical ­ presentations to

monitor: Bulbocavernosus reflex,

anal wink, external sphincter

tone, and bowel and bladder

function

HNP, herniated nucleus pulposus; TA, tibialis anterior; TFL, tensor fascia lata; TP, tibialis posterior.382 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

MUSCLES WITH DUAL INNERVATION (TABLE 5–26)

TABLE 5–26 Muscles With Dual Innervation

MUSCLE NERVES

Pectoralis major Medial pectoral Lateral pectoral nerve

Brachialis Musculocutaneous nerve Radial nerve

Flexor digitorum profundus Median nerve (AIN)—FDP 1,2 Ulnar nerve—FDP 3,4

Lumbricals Median nerve Ulnar nerve

Flexor pollicis brevis Median nerve Ulnar nerve

Pectineus Femoral nerve Obturator nerve

Adductor magnus Sciatic nerve (tibial portion) Obturator nerve

Biceps femoris Sciatic nerve (tibial portion) Sciatic nerve (peroneal portion)

AIN, anterior interosseus nerve; FDP, flexor digitorum profundus.

CHRONOLOGY OF ELECTRODIAGNOSTIC FINDINGS (TABLE 5–27)

TABLE 5–27 Chronology of Electrodiagnostic Findings

TIME ABNORMALITY

0 Decreased recruitment

Decreased recruitment interval

Prolonged F wave

Abnormal H-reflex (S1 radiculopathy)

4 days Decreased compound motor action potential amplitude (approxi-

mately 50% compared to opposite side) in severe cases

1 week Abnormal spontaneous activity occurs first in the paraspinals

They can be normal if:

•

•

They become reinnervated

The posterior primary rami are spared

They can be the only abnormal finding 10%–30% of the time

2 weeks Abnormal spontaneous activity beginning in the limbs

3 weeks Abnormal activity present in both the paraspinals and limbs

5–6 weeks Reinnervation begins

6 months to 1 year Increased amplitude from reinnervated motor unit. Reinnervation

complete

Every 3–4 months Serial EMG can be performed in intervals to monitor for reinnerva-

tion as clinically warranted

EMG, electromyography.PLEXOPATHIES

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n

PLEXOPATHIES

GENERAL

•

This is a pathologic process typically occurring distal to the DRG and proximal to the

­ peripheral nerves. Abnormalities can appear diffuse and will not follow any particular derma-

tomal or ­ myotomal distribution.

ETIOLOGY

•

Common etiologies include:

– – Trauma: Traction, transection, obstetrical injuries, compression, and hemorrhage

– – Cancer (tumor and radiation therapy)

– – Idiopathic (neuralgic amyotrophy)

ELECTRODIAGNOSTIC FINDINGS

•

Active recruitment of the weakened muscles helps to rule out more involved processes, such

as neurotmesis. However, a complete neuropraxic block can appear identical to neurotmesis or

­ axonotmesis in an acute injury if active recruitment cannot be generated.

•

NCS:

– – – – Abnormal SNAP and CMAP patterns correspond to the site of plexus injury

The SNAP help to localize if a lesion is proximal or distal to the DRG. Decreased SNAP

­ amplitudes result from axon loss distal to the DRG. If the SNAP is preserved, but there is

marked abnormality to the CMAP, then the injury is proximal.

– – The distal CMAP amplitude is the main prognostic factor in a plexopathy because it repre-

sents axonal loss in such conditions, and side-to-side comparison should be performed.

– – See specific injury patterns in this section for more details

•

Late responses:

– F-waves may be delayed or absent in plexopathies but are nonspecific, as they cannot local-

–

ize a focal lesion.

– H-reflex: Helpful to evaluate the S1 pathway but not pathognomonic

–

•

Needle EMG findings:

– – Abnormal activity in the peripheral muscles in distribution of plexus injury (e.g., upper

trunk distribution) but with normal paraspinal activity

THE BRACHIAL PLEXUS

Anatomy (Figure 5–74)

•

Origin:

– – Nerve fibers originate from the ventral rami of the C5–T1 nerve roots, which further divide

to form the trunks, division, cords, and terminal nerve branches.

•

Course:

– – The ventral rami emerge between the anterior and middle scalene muscles. In the posterior

triangle of the neck, C5 and C6 form the upper trunk, C7 forms the middle trunk, and C8

and T1 form the lower trunk.

– – The trunks pass the clavicle and form anterior and posterior divisions to become cords. The

cords are named in their relation to the axillary artery.

– – The three posterior divisions of the upper, middle, and lower trunk form the posterior cord.

The lateral cord is formed from the anterior divisions of the upper and middle trunk, and the

medial cord is formed by the anterior divisions of the lower trunk.

– The lateral cord splits to form the musculocutaneous branch and also fuses with the

–

medial cord to form the median branch. The posterior cord splits into the radial and

­ axillary branches, and the medial cord splits to contribute to the median branch and the

ulnar branch.384 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

C5 C6 C7 C8 T1

A = Anterior

division

P = Posterior

division

R

O

O

T

S

T

R

U

N

K

D

I

V

I

S

I

O

N

C

O

R

D

S

B

R

A

N

C

H

E

S

Lateral portion of

pectoralis major

(C5, C6)

Rhomboids (C5), Levator

Scapular muscle

Suprascapular nerve

Infraspinatus (C5, C6)

Supraspinatus (C4, C5, C6)

Lateral pectoral nerve

Dorsal scapular

nerve

Lateral

Posterior

Subscapularis (C5, C6, C7)

Upper subscapular nerve

A A P P A

P

Upper

CLAVI CLE

Middle

Medial

Lower

Long thoracic nerve

Serratus anterior (C5, C6, C7)

Musculocutaneous nerve

Axillary nerve

Radial nerve

Median nerve

1st

Rib

Medial pectoral nerve

Medial portion of pectoralis major (C7, C8, T1)

Pectoralis minor (C8, T1)

Medial brachial cutaneous nerve

Medial antebrachial cutaneous nerve

Ulnar nerve

Thoraco dorsal nerve

Latissimus dorsi (C6, C7, C8)

Lower subscapular nerve

Teres major (C5, C6)

Subscapularis (C5, C6, C7)

**Nerves Musculocutaneous Axillary Radial Nerve Median Nerve Ulnar Nerve**

C5 C6 • Biceps • Brachialis • Deltoid • Teres minor

• Supinator

C5 C6 C7 • Coracobrachialis • Brachioradialis

7C6C

teresrotanorP•

• Flexor carpi radialis

8C7C6C

•

ECR longus

• Triceps

8C7C

siranluipracroxelF•sugnolsiramlaP•siverbR • Ext. dig.

• EIP

• EDM

• ECU

• Abd. poll. longus

• Ext. poll. brevis

• Ext. poll. longus

CE•

1T8C7C

suenocnA•

selcsum4—SDF•

1T8C

selcsum2—PDF•selcsum2—PDF•

• FPL • Pronator quad • Lumbricals—2 muscles • Opponens pollicis • Abductor pollicis brevis • Flex poll. brevis 1/2 • Dorsal interossei—4 muscles

• Palmar interossei—3 muscles

• Lumbricals—2 muscles

• Add poll.—1 muscle

• Flex poll. brevis 1/2

• Hypothenar muscles

– Oppon. dig. min.

– Abd. dig. min.

– Flex. dig. min.

• Palmaris brevis

FIGURE 5–74 Brachial plexus. (Root levels of individual muscles are identified to the left of the table.)

ECR, extensor carpi radialis; ECU, extensor carpi ulnaris; EDM, extensor digiti minimi; EIP, ­ extensor indicis proprius;

FDP, flexor digitorum profundus; FDS, flexor digitorum supersicialis; FPL, flexor pollicis longus.PLEXOPATHIES

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FIGURE 5–75

Waiter’s tip

position: The

area of skin

that is usually

anesthetic is

shaded.

BRACHIAL PLEXUS INJURIES

Upper Trunk Brachial Plexopathy/Erb’s Palsy/“Stinger”

•

•

General: This injury involves the C5–C6 nerve roots of the upper trunk.

Etiology: It can occur from nerve traction or compression or from an obstetrical

injury. It can be sports related (stinger) and involve the C5–C6 nerve roots or

the upper trunk.

•

Clinical presentation:

– A classic manifestation is the waiter’s tip position. The arm becomes

–

adducted (deltoid and supraspinatus [SS] weakness), internally rotated

(teres minor and infraspinatus [IS] weakness), extended (bicep and

­ brachioradialis [BR] weakness), pronated (supinator and BR weakness),

with the wrist flexed (extensor carpi radialis longus [ECR-L] and brevis

­ weakness; Figure 5–75).

•

EDX pearl:

– – Stimulate at the tip of the C6 transverse process over the trunks of the bra-

chial plexus to assess Erb’s point (Figure 5–76)

•

Treatment: Rehabilitation, intermittent splinting, and activity restriction

Lower Trunk Brachial Plexopathy/Klumpke’s Palsy

•

•

General: This injury involves the C8–T1 nerve roots or lower trunk.

Etiology: It can occur from an obstetrical traction injury (Klumpke’s

palsy), forced adduction seen in an MVA, falls, shoulder disloca-

tions, Pancoast tumor, thoracic outlet syndrome (TOS), and so on.

•

Clinical presentation:

– – The patient may have wasting of the small hand muscles and

a claw hand deformity (lumbrical weakness; Figure 5–77). The

­ shoulder girdle muscle function is preserved.

•

EDX pearl:

– – The preservation of a SNAP potential may indicate a nerve root

avulsion. Avulsions may be associated with this location of injury

due to the lack of protective support at the C8 and T1 roots. In

addition, medial antebrachial cutaneous sensory response will be

absent or reduced.

•

Treatment: Rehabilitation with incomplete lesions or surgical

­ exploration with a nerve root avulsion injury

Thoracic Outlet Syndrome (TOS) (Figure 5–78)

(Lower Trunk Plexopathy)

•

•

TOS (Figure 5–78) is a type of lower trunk plexopathy due to vascular or neurogenic causes.

Vascular TOS:

– General: This injury involves the subclavian artery, subclavian vein, or axillary vein.

–

– Etiology: It can occur from a pathology resulting in arterial or venous compromise.

–

– Clinical presentation.

–

nn Arterial involvement may manifest as limb ischemia, necrosis, vague pain, or fatigue,

with decreased color and temperature. Venous involvement can manifest as a bluish,

swollen, achy limb.

•

Neurogenic TOS:

– General: This injury is rare and may actually be seen in only 1 of 1,000,000 patients.

–

– Etiology (Figure 5–78):

–

nn It can occur from compression of the lower trunk of the brachial plexus between a fibrous

band, between the first cervical rib and clavicle (costoclavicular syndrome), muscular

entrapment by the scalenes (anterior and middle scalene syndromes), or pectoralis minor

muscle (pectoralis minor syndrome).

FIGURE 5–76 Erb’s point

stimulation.

FIGURE 5–77 Ulnar claw

hand.386 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

Nerve roots

Superior trunk

Middle trunk

Inferior trunk

Lateral cord

Posterior

cord

C4

C5

C6

C7

T1

**Neurovascular and Muscular Structures**

**of the Thoracic Outlet**

Posterior scalene

Middle scalene

Anterior scalene

Brachial plexus

Inferior trunk

Middle cord

Subclavian artery

Pectoralis minor

1st rib

FIGURE 5–78 The thoracic outlet.

– Clinical presentation:

–

nn The patient may have pain and numbness along

the medial aspect of the forearm and hand, which

increases with overhead activity. Discomfort can

also be noted in the neck, clavicle, and axilla.

nn Hand muscle wasting may also be noted (median

thenar > ulnar intrinsics).

nn A maneuver to assess the neurovascular bundle is

Adson’s test (Figure 5–79). This is ­ performed by

abduction, extending and externally rotating the

patient’s arm. While ­ monitoring the radial pulse,

have the patient rotate the head toward the arm

(the side of the lesion). A decrease or loss of pulse

may be related to a compression of the subclavian

FIGURE 5–79 Adson’s test

artery, indicating compromise to the complex.

– EDX pearl:

–

nn Abnormal findings that can be noted are decreased amplitudes for the median CMAP,

ulnar SNAP/CMAP, and medial antebrachial cutaneous studies. The median SNAP is

spared. Abnormal spontaneous activity can also occur in the median and ulnar hand

muscles (lower trunk) on EMG.

– Treatment:

–

nn Rehabilitation with a focus on ROM exercises, stretching of appropriate muscles

(­ anterior/middle scalenes, pectoralis minor, trapezius and levator scapulae); strengthen-

ing of the ­ scapular stabilizers (upper/middle trapezius and rhomboids); and focus on

postural ­ mechanics to address possible entrapment syndromes against the first rib.

nn Surgery can be indicated for a first rib or fibrous band resection.

Neuralgic Amyotrophy

•

General: This injury can include various nerves of the brachial plexus.

– – Other names include Parsonage–Turner syndrome, brachial neuritis, brachial neuropathy,

­ idiopathic brachial plexopathy, shoulder-girdle neuritis, and paralytic brachial neuritis.

•

Etiology:

– – Idiopathic, inflammatory, immune-mediated process

•

Clinical presentation:

– Individuals of all ages may be affected, and there is a male predominance. The

–

most common initial symptoms are pain of abrupt onset, often severe, usuallyPLEXOPATHIES

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in the shoulder or periscapular region. It can be exacerbated by abduction and

•

•

rotation.

– – The pain may resolve in hours to days but typically begins to improve in 2 to 3 weeks, in

­ association with the development of weakness in a patchy fashion.

– – Bilateral involvement occurs in about one-third of patients, usually asymmetrically. The

patchy or multifocal involvement is common and is a hallmark of this syndrome.

EDX pearl:

– – Abnormal motor and sensory NCS and/or spontaneous activity can occur in a variety of

­ combinations: Mononeuropathy (suprascapular nerve, long thoracic nerve, axillary nerve,

­ anterior interosseus nerve [AIN], spinal accessory nerve) or plexopathy. Muscles innervated

by the C5 and C6 levels are more commonly affected.

Treatment:

– – Interventions may include rehabilitation to prevent contractures. Neuralgic amyotrophy can

resolve spontaneously.

Neoplastic Versus Radiation Plexopathy (Table 5–28)

•

General:

– – Brachial plexopathies can be caused by compression from tumors and/or from their

treatments.

– – A primary plexus tumor can arise from schwannomas, neuromas, and neurofibromas.

– – Secondary plexus involvement can arise from a Pancoast tumor from the lung or breast.

– – Radiation therapy can cause neural fibrosis and constriction of the vasa nervorum, leading

to destruction of the axon and Schwann cells. This can occur months or years after radiation

­ treatment.

•

Clinical presentation/EDX pearl:

TABLE 5–28 Neoplastic Versus Radiation Plexopathy

EMG, electromyography.

Nerve Root Avulsion (Figure 5–80)

•

General:

– – A severe plexus injury can lead to disruption of the nerve roots. This can occur from a

­ traction injury that disrupts the protective connective tissue support. The C8 and T1 roots

have less ­ protection and are the most common site of tearing from spinal cord attachments.

MRI is valuable in differentiating between a nerve root stretch and an avulsion.

•

Clinical presentation:

– – The patient complains of absent sensation or muscle contraction from the muscles inner-

vated by the roots involved. This may manifest as a flail shoulder.

•

EDX pearl:

– Absent CMAPs with normal SNAPs. Needle EMG reveals absent recruitment and

–

­ abnormal spontaneous activity in a myotomal distribution of the avulsed nerve root, includ-

ing the paraspinals.

LUMBOSACRAL PLEXUS (FIGURE 5–81)

Anatomy

•

Origin:

– Nerve fibers originating from the ventral rami of L1, L2, L3, and L4 roots form the lumbar plexus.

–

– – Nerve fibers originating from the ventral rami of L4, L5, S1, S2, S3, and S4 roots form the

sacral plexus.

CHARACTERISTICS RADIATION TUMOR

Common site of injury Upper trunk Lower trunk

Clinical presentations Myokymia on EMG Horner’s syndrome

Sensation Painless Painful388 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

**A**

**B**

Dura

Intervertebral

foramen

Nerve

root

Connective

tissue moorings

**C**

**D**

FIGURE 5–80 Nerve

root avulsion.

(A) Normal nerve

root exiting the

intervertebral

foramen. (B) Slight

traction with

dura plugging

the foramen

and support by

tissue moorings.

(C) Ruptured dura

and moorings.

(D) Ruptured roots.

Note: Arrows depict

direction of traction

forces.

•

Course:

– – – – The ventral rami then divide to form anterior and posterior divisions in each plexus.

The anterior division of the lumbar plexus forms the obturator nerve, while the posterior

division forms the femoral nerve and the lateral femoral cutaneous nerve.

– – Terminal branches directly off the lumbar plexus include:

nn Iliohypogastric nerve

nn Ilioinguinal nerve

nn Genitofemoral nerve

– In the sacral plexus, the tibial portion originates from the anterior division, while the

–

­ common peroneal nerve originates from the posterior division. The lumbosacral trunk

forms from the L4 and L5 nerve fibers connecting the lumbar to the sacral plexus and

travels over the pelvic brim.

– – Terminal branches directly off the sacral plexus include:

nn Superior gluteal nerve

nn Inferior gluteal nerve

LUMBOSACRAL PLEXOPATHIES

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Multiple potential etiologies:

– – Neuralgic amyotrophy: Similar to brachial plexus pathology

– – Neoplastic versus radiation plexopathy: Similar to brachial plexus pathology

– – Retroperitoneal bleed: This can involve a hematoma formation in the psoas muscle.

– – Hip dislocation

– – Obstetric injuries/cephalopelvic disproportion: Presents as a postpartum foot drop

•

EDX pearl:

– – Lumbar plexus evaluation:

nn SNAP: Lateral femoral cutaneous nerve (L2–L3), saphenous nerve (L4)

nn CMAP: Femoral nerve (L2–L4)

nn EMG: Muscles innervated by the femoral nerve, obturator nerve, and the iliopsoas

muscle. Normal paraspinals.

– – Sacral plexus evaluation:

nn nn nn SNAP: Superficial peroneal nerve (L5), sural nerve (S1)

CMAP: Deep peroneal nerve (L4–S1), tibial nerve (L5–S2)

EMG: Muscles innervated by the tibial nerve, peroneal (fibular) nerve, and superior and

­ inferior gluteal nerves. Normal paraspinals.PLEXOPATHIES

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L5 S1 S2

S3

L4

L2 L3

Lateral femoral cutaneous nerve

Superior gluteal nerve

Tensor fascia lata (L4, L5)

Gluteus medius (L4, L5, S1)

Gluteus minimus (L4, L5, S1)

Inferior gluteal nerve

Gluteus maximus

(L5, S1, S2)

Sciatic nerve

Saphenous

nerve

Peroneal division

Biceps femoris

(Short head)

(L5, S1, S2)

Tibial division

Semimembranosus

(L5, S1, S2)

Semitendinosus

(L5, S1, S2)

Biceps femoris (Long

head) (L5, S1, S2)

Adductor magnus (L4)

Femoral nerve

Sartorius (L2, L3)

Iliacus (L2, L3)

Pectineus (L2, L3)

Quadriceps femoris

Vastus medialis (L2, L3, L4)

Vastus intermedius (L2, L3, L4)

Vastus lateralis (L2, L3, L4)

Rectus femoris (L2, L3, L4)

Obturator nerve

Obturator externus (L3, L4)

Pectinius (L2, L3 - may receive a

branch from the obturator nerve

Adductor brevis (L2, L3, L4)

Adductor longus (L2, L3, L4)

Gracilis (L2, L3)

Adductor magnus

Sural

nerve

Common peroneal nerve

Deep peroneal nerve

Extensor digitorum longus (L5, S1)

Tibialis anterior (L4, L5)

Extensor hallucis longus (L5, S1)

Peroneus tertius (L5, S1)

Extensor digitorum brevis (L5, S1)

Superficial peroneal nerve

Peroneus longus (L5, S1)

Peroneus brevis (L5, S1)

Tibial nerve

Gastocnemius (Medial head) (S1, S2)

Gastocnemius (Lateral head) (S1, S2)

Plantaris (S1, S2)

Soleus (S1, S2)

Popliteus (L4, L5, S1)

Posterior tibial nerve

Tibialis posterior – four digits (L4, L5)

Flexor digitorum longus (S2, S3)

Flexor hallucis longus (S2, S3)

Medial plantar nerve

Flexor digitorum brevis (S2, S3)

Abductor hallucis (S2, S3)

Flexor hallucis brevis (S2, S3)

First lumbrical (S2, S3)

Lateral plantar nerve

Abductor digiti minimi (S2, S3)

Quadratus plantae (S2, S3)

Flexor digiti minimi brevis (S2, S3)

2-4 Lumbricals – three muscles (S2, S3)

Dorsal interossei – four muscles (S2, S3)

Plantar interossei – three muscles (S2, S3)

Abductor hallucis (S2, S3)

FIGURE 5–81 The lumbosacral plexus and innervations.390 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

n

UPPER LIMB MONONEUROPATHIES

MEDIAN NERVE

Anatomy

•

Origin: (Figure 5–82)

– – Nerve fibers from the C5–T1 nerve roots contribute to the upper, middle, and lower trunks

® medial and lateral cords ® median nerve.

•

Course.

ARM

•

The nerve runs medial to the axillary artery. It continues down the humerus and runs under the

­ ligament of Struthers (LOS) at the medial epicondyle of the humerus.

FOREARM

•

Innervations and cutaneous branches:

– – Pronator teres (PT)

– – FCR

– – Palmaris longus

– – Flexor digitorum superficialis (FDS)

– – Palmar cutaneous branch

– – The AIN branches from the median nerve to innervate (four Ps):

nn Flexor pollicis longus (FPL)

nn Flexor digitorum profundus (FDP 1 and 2)

nn Pronator quadratus (PQ)

HAND

•

Through the carpal tunnel, the median nerve innervates the “LOAF” muscles:

– – Lumbricals (1, 2)

– – Opponens pollicis

Median n.

Pronator teres n.

Flexor digitorum

sublimis n.

Flexor pollicis

longus n.

Flexor digitorum

profundus n.

Pronator

quadratus n.

Abductor

pollicis brevis n.

Opponens

pollicis n.

Superfic. head

of flexor pollicis

brevis n.

1st and 2nd

lumbricals

Flexor carpi radialis n.

Palmaris longus n.

Flexor digitorum

profundus n.

Anterior interosseous n.

**Cutaneous Innervation**

1

2

Posterior Anterior

FIGURE 5–82 The median n., nerve. Cutaneous

branches in hand supply: (1) Thenar eminence.

(2) Palmar surface as indicated.

n., nerve.UPPER LIMB MONONEUROPATHIES

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Brachial

artery

Supracondylar

process

Ligament of

Struthers

Median

nerve

Medial

epicondyle

FIGURE 5–83 The ligament of Struthers.

– – Abductor pollicis brevis

– – Flexor pollicis brevis (superficial)

– – (Digital cutaneous branches)

Injury

ARM

LIGAMENT OF STRUTHERS (FIGURE 5–83)

•

General:

– – There is a 2 cm bone spur 3 to 6 cm proximal to the medial epicondyle. It is connected to the

­ epicondyle by a ligament in 1% of the population.

•

Etiology:

– – The nerve becomes entrapped with the brachial

artery under the ligament.

•

Clinical presentation:

– The patient may have involvement of all median

–

nerve innervated muscles. It can manifest as ­ weakness

in grip strength (FDP and FDS weakness) and wrist

flexion (FCR weakness). A dull, achy sensation can

occur in the distal forearm. There may be difficulty

in flexing the second and third digits (FDP weak-

ness) resulting in an active benediction sign. The

brachial pulse is possibly diminished.

•

EDX findings:

– NCS: Abnormal median SNAP and CMAP

–

– EMG: Abnormal in median nerve-innervated

–

muscles, including the PT

•

Treatment:

– – Rehabilitation or surgical release

BICIPITAL APONEUROSIS (LACERTUS FIBROSUS) (FIGURE 5–84)

•

General:

– This is a thickening of the antebrachial fascia attaching the biceps to the ulna. It overlies the

–

median nerve in the proximal forearm.

•

Etiology:

– – The nerve can be injured by entrapment

or hematoma compression resulting from

an arterial blood gas or venipuncture.

•

Clinical presentation:

•

•

– – – – – – This is similar to the LOS pathology.

EDX findings:

This is similar to the LOS pathology.

Treatment:

Interventions may include rehabilitation

or surgical release.

FOREARM

PRONATOR TERES SYNDROME (FIGURE 5–85)

•

General:

– – The median nerve passes between the

two heads of the PT muscle and continues

down under the FDS muscle. An entrap-

ment is commonly named for the muscle

compressing the nerve. That muscle is

usually spared because it receives its

innervation before it is pierced by the

nerve. However, the nerve can also con-

tribute to the innervations as it courses

through the muscle.

**Lateral**

**Medial**

Biceps

brachii

Median

nerve

Brachioradialis

Bicipital

aponeurosis

(lacertus

fibrosus)

Sublimis

arch

FIGURE 5–84 The lacertus fibrosus.392 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

Median

nerve

Pronator

teres

(humeral

head)

Pronator

teres

(ulnar

head)

•

•

•

•

Etiology:

– – The nerve can be injured by compression between

the heads of the PT muscle or the bridging fascial

band of the FDS muscle.

Clinical presentation:

– – The muscles involved include all median inner-

vated muscles except the PT. The patient may

complain of a dull ache of the proximal forearm

exacerbated by forceful pronation (PT) or finger

flexion (FDS). The forearm and hand muscles may

become easily fatigued.

EDX findings:

– NCS: Abnormal median nerve SNAPs and CMAPs

–

– EMG: Abnormal activity in all median nerve inner-

–

vated muscles EXCEPT the PT

Treatment:

– – Interventions may include rehabilitation or surgical

release.

ANTERIOR INTEROSSEUS NERVE (AIN) SYNDROME

•

General:

– – This is an injury to a motor nerve branch of the

median nerve. It is a pure motor syndrome. It lacks

cutaneous innervations but has sensory branches to

the wrist joint.

– It supplies the FPL, PQ, and FDP 1, 2 (the four P

–

muscles). The FPL is usually the first muscle affected.

•

Etiology:

– – The nerve can be injured by an idiopathic process,

fracture of the forearm, lacerations, or compression.

FIGURE 5–85 Median nerve entrapment

at the pronator teres or flexor digitorum

superficialis (flexor sublimis).

•

Clinical presentation:

– – The patient may demonstrate a positive (abnormal) “OK” sign (Figure 5–86) or have dif-

ficulty forming a fist (Figure 5–87) because of an inability to approximate the thumb and

index finger (FPL, FDP weakness). The muscles involved include FPL, PQ, and first and

second heads of the FDP (the four P muscles). Sensation is spared.

•

EDX findings:

– NCS: Normal median nerve SNAP and median nerve CMAP to the abductor pollicis brevis (APB).

–

Possible abnormal CMAP to the PQ.

– EMG: Abnormal activity in the muscles innervated by the AIN (FPL, FDP 1 and 2, and PQ)

–

•

Treatment:

– – Interventions may include rehabilitation or surgical exploration.

Sublimis

bridge

Flexor

digitorum

superficialis

A B

A B

FIGURE 5–86 (A) Normal. (B) Positive OK sign

(or pinch sign).

FIGURE 5–87 (A) Normal. (B) Inability to make a fist due to

problems incorporating the thumb and index finger.UPPER LIMB MONONEUROPATHIES

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•

CARPAL TUNNEL SYNDROME

General:

– – This is a median neuropathy caused by compression of the median nerve in the carpal tun-

nel and is the most common nerve entrapment in the upper extremity.

– – It is characterized by a clinical syndrome in which nerve damage is coupled with abnormal

­ sensations and pain. It can involve the first, second, third, and 1/2 of the fourth digit.

– – The EDX abnormality alone should not establish the diagnosis, as 10% to 15% of people with

clinical carpal tunnel syndrome (CTS) can have a normal study.

CARPAL TUNNEL CONTENTS (FIGURE 5–88)

FCR, flexor carpi radialis; FDP, flexor digitorum profundus; FDS, flexor digitorum superficialis.

CONTENTS: NINE TENDONS AND ONE

NERVE BORDERS

Superficial layer

Four FDS tendons

One FPL tendon

Median nerve

(FCR is outside the carpal tunnel)

Transverse carpal ligament

Carpal arch bones

Deep layer

Four FDP tendons

FDS (Flexor digitorum

superficialis m.) (tendons)

FDP (Flexor digitorum

profundus m.) (tendons)

Median nerve

Transverse carpal ligament

FCR (Flexor carpi

radialis m.) (tendons)

FPL (Flexor pollicis

longus m.) (tendons)

Carpal bones

FIGURE 5–88 Cross-section of the carpal tunnel (at level of the first row of the carpal bones).

•

•

Etiology:

– – The nerve can be injured in the carpal tunnel by:

nn Idiopathic process

nn Increased canal volume from thyroid disease, congestive heart failure (CHF), renal ­ failure,

mass (tumor, hematoma), and pregnancy (it usually occurs at 6 months and resolves

postpartum).

nn Decreased canal volume from a fracture, arthritis, and rheumatoid tenosynovitis.

nn Double crush syndrome from diabetes mellitus, cervical radiculopathy, and TOS.

Clinical presentation:

– – Symptoms usually begin gradually, and are often experienced at night. The thumb, index,

and middle fingers are most frequently affected. The most common symptoms of CTS

include night numbness, tingling, and pain in the hand; a feeling of electric shock in the

fingers or lateral hand.

– – Sensation is abnormal to the lateral 3–1/2 fingers of the hand except at the base of the

thumb. Muscle weakness can be noted in “LOAF” muscles (Lumbricals 1 and 2, Opponens

pollicis, Abductor pollicis brevis, and Flexor pollicis brevis).

– – Provocative maneuvers are noted in Table 5–29PROVOCATIVE TESTS DESCRIPTION

Tinel’s sign Percussion of the median nerve at the wrist

Phalen’s test Hold the wrist at 90 degree flexion for approximately 1 minute

Tourniquet test Inflated BP cuff reproduction of symptoms at 1 minute

Carpal compression test Hold thumb compression over the tunnel for 30 seconds

Reverse Phalen’s test Hold the wrist at 90 degree of extension for approximately 1 minute

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– – The severity of clinical symptoms from CTS does not necessarily correlate with severity of

EDX findings, but in general the clinical presentation is as follows:

•

Mild CTS:

– – The patient may complain of numbness, paresthesias, or dysesthesias radiating to the first,

second, third, and lateral fourth digits. Symptoms may be exacerbated during sleep and

relieved with wrist shaking.

•

Moderate CTS:

– The patient may complain of continuous sensory deficits in the median nerve distribution,

–

­ involving the entire palm and radiating proximally . The ability to handle fine objects is impaired.

•

Severe CTS:

– The patient may complain of severe sensory loss and muscular atrophy of the thenar eminence.

–

TABLE 5–29 Provocative Tests for Carpal Tunnel Syndrome

BP, blood pressure.

•

•

EDX findings (Table 5–30):

– – Sensory studies are more sensitive than motor studies because larger sensory fibers are more

affected than the slightly smaller motor nerve fibers.

– – Antidromic sensory studies are typically performed and offer the advantage of producing

larger amplitudes and are more commonly practiced.

Special studies:

– On routine median NCSs, a demyelinating lesion at the carpal tunnel results in slowing

–

and ­ prolongation of the distal motor and sensory latencies. If the demyelination has resulted

in ­ conduction block or secondary axonal loss, the distal CMAP and SNAP amplitudes will be

decreased as well. The study of distal segments of the median nerve is helpful in distinguish-

ing CTS from peripheral neuropathy; in CTS, the maximal slowing is across the wrist, whereas,

in peripheral neuropathy, the distal segment is more abnormal.

– Approximately 10% to 25% of CTS patients have normal routine median conduction studies;

–

in such patients, further testing is performed using more sensitive NCSs. Those studies usu-

ally involve a comparison of the median nerve to ulnar nerve in the same hand. The common

median-versus-ulnar comparison tests are (a) median-­ versus-ulnar palm-to-wrist mixed-nerve

latencies (the typical cutoff is 0.4 msec), and (b) median-­ versus-ulnar wrist-to-digit-4 sensory

latencies (a difference of 0.5 msec is considered abnormal).

TABLE 5–30 EDX Findings for Carpal Tunnel Syndrome

CMAP, compound motor action potential; EMG, electromyography; NCS, nerve conduction studies; SNAP, sensory

nerve action potential.

SEVERITY NCS EMG

Mild SNAP: Prolonged latency

CMAP: Normal

Normal

Moderate SNAP: As above plus decreased amplitude

CMAP: Prolonged latency

Normal

Severe SNAP: Absent

CMAP: As above plus decreased amplitude

Abnormal activityUPPER LIMB MONONEUROPATHIES

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Median

nerve

•

•

Treatment:

Interventions may include the following:

– Conservative

–

nn Indications: Mild symptoms with no weakness/atrophy or denervation on EMG.

nn Orthotics: Hand splint neutral to 30-degree extension.

nn Medications: Nonsteroidal anti-inflammatory drugs (NSAIDs), or a steroid injection,

diuretics, and vitamin B6.

nn Ergonomic modifications

nn Treat underlying medical disorders

– Surgical indications:

–

nn Persistent numbness/paresthesias, severe pain despite a trial of conservative treatment.

nn Severe CTS with severe muscle atrophy typically does not have a high surgical success

rate due to the degree of the nerve damage that has already occurred.

Prognosis:

– – Poor outcome with conservative management may occur with:

nn Symptoms >10 months in duration

nn Constant paresthesias

nn Positive Phalen’s test in <10 seconds

nn Weakness, atrophy

nn Marked prolonged latency on NCS

nn Abnormal spontaneous activity on EMG

ANOMALOUS INNERVATIONS

MARTIN-GRUBER ANASTOMOSIS (FIGURE 5–89)

•

General:

– This is a median to ulnar nerve anastomosis. Fibers

–

from the AIN branch of the median nerve ­ anastomose

with the ulnar nerve. Alternatively, nerve fibers of the

proximal median nerve cross over to the ulnar nerve in

the forearm to innervate the adductor pollicis (ADP),

abductor digiti minimi (ADM), and most commonly the

first dorsal interossei (DI) ­ muscles. This can occur in 15%

to 20% of the population.

•

EDX findings (Figure 5–90):

1. An initial positive deflection of the median nerve

CMAP (Figure 5–90A) with median nerve stimulation at

the antecubital fossa, which is not seen at the wrist. This is

due to the proximal stimulus stimulating the ulnar nerve/

muscles through the anastomosis, and the potential from

the ulnar muscle fibers reaching the recording electrode

via volume conduction before the ­ potential from the

median nerve, as a result of the median nerve fibers being

entrapped and slowed down at the wrist.

2. An increased amplitude of the median nerve CMAP

(Figure 5–90B) is seen at the elbow compared to the wrist.

FIGURE 5–89 Martin-Gruber anastomosis.

This is due to simultaneously stimulating the median and the ulnar nerve innervated muscles at

the elbow. This can occur without a median nerve entrapment.

3. An artificially fast CV can be demonstrated. This is a mathematical error due to using a

normal ­ proximal latency reading obtained by ­ stimulating the branch to the ADM. This is

subsequently ­ calculated with the prolonged distal latency obtained by the entrapped median

nerve, giving a falsely increased CV .

Ulnar

nerve

Martin-Gruber

anastomosis

Example

Elbow latency (6.0 msec) − Wrist latency (4.5 msec) = 1.5 msec

Forearm measurement (150 mm)/Forearm latency (1.5 msec) = 100 m/sec396 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

FIGURE 5–90 Martin-Gruber

anastomosis with carpal tunnel

syndrome electrodiagnostic

finding: (A) Initial positive

deflection of the CMAP is caused

by response of ulnar muscles

innervated by the median nerve,

which has gotten to the recording

electrode earlier, since the aberrant

portion of the median nerve is not

entrapped. (B) Increased amplitude

of the median nerve CMAP is seen

at the elbow (as opposed to the

wrist). This is due to simultaneously

stimulating the median and ulnar

innervated muscles at the elbow.

CMAP, compound motor action

potential.

5

6

7

1

2

Ulnar nerve

Med. cut. n. of arm

Med. cut. n. of

forearm

Cutaneous

distribution

•

•

RICHE-CANNIEU ANASTOMOSIS

General:

– This is a connection of the recurrent branch of the median nerve in the hand to the deep motor

–

branch of the ulnar nerve. It produces an all ulnar innervated hand.

EDX findings:

– While recording over the APB muscle, the median nerve CMAP is absent with median nerve

–

stimulation but present with stimulation of the ulnar nerve.

ULNAR NERVE

Anatomy

•

Origin (Figure 5–91):

– – These nerve fibers originate from the C8–T1

roots. They continue on to ­ contribute to the

lower trunk, medial cord, and finally form the

ulnar nerve.

•

Course:

ARM

•

The nerve descends along the medial surface of

the medial head of the triceps. It runs within a

deep groove of thick fascia known as the arcade of

Struthers (AOS). It continues posteriorly in a sulcus

between the medial epicondyle and olecranon

called the retrocondylar groove.

FOREARM

•

It then continues distally in the cubital ­ tunnel to

innervate the:

– – Flexor carpi ulnaris (FCU)

– – FDP (third and fourth)

– – Palmar ulnar cutaneous nerve

– Dorsal ulnar cutaneous (DUC) nerve: This

–

branch does not travel through Guyon’s canal

(­ arising 5–8 cm more proximally) and is nor-

mal in a distal ulnar neuropathy at the wrist

but abnormal in more proximal compressions.

– – Dorsal digital nerves

FIGURE 5–91 The ulnar nerve.

Flexor carpi

ulnaris

Flexor

digitorum

profundus

Adductor

pollicis

Anterior

Posterior

2

1

2

3 4

Palmaris brevis

Abductor

Opponens

Flexor

Digiti

quinti

3rd & 4th lumbricals

Palmar & dorsal

interossei

1

3UPPER LIMB MONONEUROPATHIES

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The 1-1/2 Nerve

•

1-1/2 muscles of the thumb

•

1 Adductor pollicis

•

1/2 Flexor pollicis brevis (deep head)

•

1-1/2 muscle of the forearm

•

1 Flexor carpi ulnaris

•

1/2 Flexor digitorum profundus (3, 4)

•

1-1/2 sensation of the fingers

•

1 Fifth digit

•

1/2 Fourth digit

HAND

•

Through Guyon’s canal, it splits into three branches:

– Superficial sensory branch.

–

– Hypothenar branch:

–

nn Opponens digiti quinti

nn Abductor digiti quinti

nn Flexor digiti quinti

– Deep motor branch:

–

nn Palmaris brevis

nn 4 Dorsal interossei—(“DAB”: Abduction)

nn 3 Palmar interossei—(“PAD”: Adduction)

nn 2 Lumbricals

nn 1 Adductor pollicis

nn 1/2 Flexor pollicis brevis (deep head)

Injury

ARM

ARCADE OF STRUTHERS (FIGURE 5–92)

•

General:

– – This is a fascial band in the medial arm that connects the brachialis to the triceps brachii.

•

Etiology:

– – The ulnar nerve can be injured due to compression under the fascial band.

•

Clinical presentation:

– – The patient may demonstrate involvement of all ulnar nerve innervated muscles.

– – Wrist flexion may result with a radial deviation (FCU weakness).

– – Abnormal sensations may occur in all sensory branches of the ulnar nerve.

– Ulnar claw hand (Figure 5–93): While the hand is at rest, an unopposed pull of the extensor

–

digitorum ­ communis (EDC) causes partial finger flexion of the fourth and fifth PIP and DIP

joint due to extension of the MCP .

Biceps

Coracobrachialis

Ulnar nerve

(anteriorly)

Brachialis

Medial

intermuscular

septum

Ulnar nerve

(posteriorly)

Medial

head of

triceps

Arcade of

Struthers

FIGURE 5–92 The Arcade of Struthers. •

•

•

Special signs:

– Froment’s sign (Figure 5–94): This test demon-

–

strates an inability to hold a piece of paper by the

thumb and index finger with pure thumb adduc-

tion (adductor pollicis weakness). The patient

substitutes the median innervated FPL muscle,

causing flexion of the ­ interphalangeal joint.

– Wartenberg’s sign: Inability to adduct the fifth

–

digit (interossei weakness).

EDX findings:

– NCS: Abnormal DUC SNAP; abnormal ulnar

–

SNAP and CMAP

– EMG: Abnormal activity in all the ulnar nerve

–

innervated muscles

Treatment: Interventions may include

­ rehabilitation or surgical release of the AOS.

FIGURE 5–93 Ulnar claw hand.

FIGURE 5–94 (A) Normal. (B) Froment’s sign.5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

•

•

•

•

Ulnar

nerve

Medial

epicondyle

Cubital tunnel

Olecranon

398 FOREARM

TARDY ULNAR NERVE PALSY

•

General:

– – This is an ulnar neuropathy that can occur months to years after a distal humeral fracture.

•

Etiology:

– – The nerve can be injured secondary to a trauma that results in bone overgrowth or scar

­ formation. Nerve traction can also occur from an increased carrying angle due to a valgus

deformity at the elbow.

Clinical presentation:

– The patient’s muscle involvement and complaints are dependent on the site of injury. In most

–

cases, the patient may demonstrate involvement of all the ulnar nerve innervated muscles.

EDX findings:

– – Abnormal ulnar SNAP and CMAP and abnormal EMG of all ulnar muscles

Treatment:

– – Interventions may include rehabilitation or surgical procedure.

CUBITAL TUNNEL SYNDROME

General:

– – This is the most common site of

elbow entrapment.

– It is bordered by the medial ­ epicondyle

–

and olecranon with an overlying apo-

neurotic band. NCS across the elbow

should be ­ performed at 90-degree to

110-degree elbow flexion (Figure 5–95).

This will avoid underestimation of

actual nerve length, thus causing false

positive findings.

Etiology:

– – The nerve can be injured from

­ compression beneath the proximal

edge of the FCU aponeurosis or arcu-

ate ligament.

Clinical presentation:

The muscles involved include all ulnar nerve innervated muscles with one exception: The

FCU may or may not be involved. The patient may complain of symptoms similar to an injury

at the AOS. A ­ positive Tinel’s sign may demonstrate paresthesias in the distribution of the

ulnar nerve with percussion at the ulnar groove.

EDX findings:

– NCS: Motor conduction studies are more useful than sensory NCS. Recording is typically at

–

the ADM. However, the first DI may show earlier abnormalities due to those nerve fascicles

being more prone to injury.

– – A decrease in ulnar CMAP amplitude seen both below and above the elbow may be second-

ary to a Martin-Gruber anastomosis. This can be checked by stimulating the median nerve

at the elbow, which will provide a waveform with an amplitude equal to what is considered

“missing” from the ulnar nerve stimulation.

nn SNAP: Abnormal ulnar nerve and DUC findings

nn CMAP: Approximately 10 to 15 msec decrease in CV across the elbow (most consistent

­ finding); a 20% drop in amplitude is abnormal

– EMG:

–

nn Abnormal activity in the ulnar nerve innervated (hand intrinsics > forearm) muscles. Due

to nerve fascicle placement in the retrocondylar groove there is a 70% chance of find-

ing abnormalities in the hand intrinsics compared to a 25% chance in the ulnar forearm

muscles. Furthermore, the FDP has more fascicles that pass through the cubital tunnel

and is more ­ reliable to test than the FCU.

Treatment:

– – Interventions may include rehabilitation or surgical release.

•

Flexor

carpi ulnaris

FIGURE 5–95 The cubital tunnel.

•

– – •

•UPPER LIMB MONONEUROPATHIES

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HAND

GUYON’S CANAL

•

General:

– Different branches of the ulnar nerve can be injured at the wrist (Figure 5–96).

–

•

Etiology:

SHEA’S CLASSIFICATION SYSTEM OF ULNAR INJURIES AT GUYON’S CANAL

Type I Involvement of the deep ulnar branch, hypothenar, and sensory

Type II Involvement of deep ulnar motor branches

Type III Involvement of the superficial ulnar sensory branch

Note: There can also be involvement of the ulnar nerve proximal to or at Guyon’s canal to involve all three branches:

the ­ hypothenar, deep ulnar motor, and superficial ulnar sensory branches.

•

•

•

– – The nerve can be injured by cycling activities (cyclist’s palsy), wrist ganglions, or rheuma-

toid arthritis (RA), trauma, or carpal bone fracture.

Clinical presentation:

– – The muscles involved include all the ulnar nerve innervated intrinsic muscles of the hand

(see box on top of next page).

– – The patient may complain of painless wasting of the first DI. A severe claw hand may occur

(­ lumbrical weakness) while the FDP remains intact, causing marked finger flexion.

EDX findings:

– NCS:

–

nn SNAP: Ulnar SNAP to the fifth digit is abnormal, but DUC nerve SNAP is spared, as the

DUC nerve is proximal to lesion.

nn CMAP: Abnormal (depends on which branch[es] affected)

– EMG: Abnormal activity in the ulnar nerve innervated hand muscles

–

Treatment:

– – Interventions may include rehabilitation or a surgical procedure.

Dorsal ulnar

cutaneous

sensory

Palmar

cutaneous

sensory

Hypothenar

motor

Digital

sensory

3

Deep palmar

motor branch

Pisiform

bone

4 4

2

1

Hook of

the hamate

FIGURE 5–96 Detailed anatomy of the ulnar nerve at the wrist. Entrapment of the ulnar nerve at the wrist can take

on several patterns: (1) pure motor affecting only the deep palmar motor branch, (2) pure motor affecting the deep

palmar and hypothenar motor branches, (3) motor and sensory (proximal canal lesion), and rarely (4) pure sensory

involving only the sensory fibers to the volar fourth and fifth fingers.400 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

Muscles Involved

4 Dorsal interossei

3 Palmar interossei and the Hypothenar eminence:

2 Lumbricals

•

Opponens digiti minimi

1 Adductor pollicis

•

Abductor digiti minimi

1/2 Flexor pollicis brevis

•

Flexor digiti minimi

Note: Compression of the ulnar nerve at Guyon’s canal may spare the hypothenar muscle.

RADIAL NERVE

Anatomy

•

Origin (Figure 5–97):

– – These nerve fibers originate from the C5–T1 roots. They continue on to contribute to the

upper, middle, and lower trunks, posterior cord, and finally form the radial nerve.

•

Course:

C5

Radial nerve

Post. cut. nerve

of arm

Lower lat.cut.

nerve of arm

Post. cut. nerve

of forearm

5

6

7

1

2

Triceps

Triceps & anconeus

Brachioradialis

Extensor carpi radialis longus

Posterior interosseus nerve

Extensor carpi radialis brevis

Supinator

Extensor digitorum

Extensor digiti quinti

Extensor carpi ulnaris

Abductor pollicis

longus

Extensor pollicis

longus & brevis

Extensor indicis

Cutaneous

innervation

Dorsal digital n’s

Anterior

Posterior

FIGURE 5–97 The radial nerve.

ARM

•

The nerve is located posterior to the axillary artery. It descends between the long and medial

heads of the triceps muscle toward the spiral groove.

– – Muscles innervated above the spiral groove:

nn Triceps brachii

nn Anconeus

nn Posterior cutaneous nerve

nn Lower lateral cutaneous nerveUPPER LIMB MONONEUROPATHIES

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– Muscles innervated below the spiral groove:

–

•

nn BR

nn ECR-L

nn Posterior cutaneous nerve of forearm

10 cm proximal to the lateral epicondyle of the humerus, the nerve pierces the lateral intermuscular

septum and enters the anterior compartment of the arm. It continues distally between the brachio-

radialis and brachialis.

At the lateral epicondyle it then splits into a motor (posterior interosseus nerve [PIN]) and

sensory branch (superficial radial nerve)

ELBOW

•

Superficial radial nerve

•

Posterior Interosseus Nerve innervates:

– – Extensor carpi radialis brevis (ECR-B)

– – Supinator

– – EDC

– – Extensor digiti minimi (EDM)

– – Extensor carpi ulnaris (ECU)

– – Abductor pollicis longus (APL)

– – Extensor pollicis longus (EPL)

– – Extensor pollicis brevis (EPB)

– – Extensor indicis proprius (EIP)

Injuries

AXILLA

CRUTCH PALSY

•

General:

– – Nerve injury at this level can involve the posterior cord of the brachial plexus, most com-

monly affecting the radial nerve.

•

Etiology:

– – The radial nerve or posterior cords of the brachial plexus can become compressed with

improper axillary crutch use.

•

Clinical presentation:

– – The patient may complain of weakness in all radial nerve innervated muscles, including the

­ triceps brachii. Sensation may be decreased over the posterior arm and forearm.

•

EDX findings:

– NCS:

–

nn Abnormal radial SNAP and CMAPs

nn Injury to the posterior cords of the brachial plexus, which can affect the radial, axillary, and/

or suprascapular nerves, can result in abnormal SNAPs and CMAPs of the affected nerves.

– EMG:

–

nn Abnormal activity in all radial nerve innervated muscles

nn Injury to the posterior cords will result in abnormal activity in corresponding muscles

that are innervated by the injured posterior cords.

•

Treatment:

– – Interventions may focus on rehabilitation, discontinuing

crutch use, static cock up splint, or dynamic splinting

ARM

SPIRAL GROOVE

•

General:

– – Injury at this site is also known as Saturday night palsy or

honeymooner’s palsy.

•

Etiology:

– The most common cause of radial nerve injury at the spiral

–

groove is trauma, notably with humeral fractures.

FIGURE 5–98 Mechanism of

honeymooner’s palsy (also known

as Saturday night palsy).402 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

•

•

•

– – Other common mechanisms of radial nerve injury at the spiral groove include iatrogenic

injury (upper limb surgery, BP cuff used during surgery), compression (“Saturday night

palsy” (Figure 5–98)), and IM injection.

Clinical presentation:

– – Weakness noted in radial nerve innervated muscles below the spiral groove. Triceps brachii

and anconeus are spared due to innervation above the spiral groove.

– – The patient complains of weakness of elbow flexion (BR weakness), supination (supinator

­ weakness), wrist drop (ECR-L, ECR-B, ECU weakness), and finger extension (EDC weak-

ness). There is preservation of elbow extension (triceps, anconeus). Sensory deficits may

occur in the dorsal aspect of the hand and posterior forearm.

EDX findings:

– NCS: Abnormal radial SNAPs and CMAP

–

– EMG: Abnormal activity in all radial nerve innervated muscles below the spiral groove. The

–

­ triceps is innervated proximal to the spiral groove, so the triceps should be normal.

Treatment:

– – Intervention is primarily focused on rehabilitation. Surgery is indicated in cases of nerve

­ transection, fractures, or open injuries.

Differential Diagnosis of a Wrist Drop

Mononeuropathy: PIN, radial nerve

Radiculopathy: C6 or C7

Diffuse polyneuropathy: Lead

Plexopathy: Posterior cord, upper trunk, middle trunk

Central: SCI/TBI/CVA, etc.

CVA, cerebrovascular accident; PIN, posterior interosseous nerve; SCI, spinal cord injury; TBI, traumatic brain injury.

FOREARM

RADIAL TUNNEL SYNDROME

•

General:

– – The anatomical course of the radial nerve at the elbow is highly variable. It typically runs

through an intramuscular septum between the brachialis and BR. Branching of the radial

nerve into the PIN and superficial radial branches can occur before or after it travels through

the intramuscular septum.

•

Etiology:

– The radial nerve can be entrapped between the brachialis and BR in the radial tunnel in the

–

elbow.

•

Clinical presentation:

– – Patients typically present with lateral proximal forearm pain that worsens with activity,

which can mimic lateral epicondylitis.

– Patients with radial tunnel syndrome will present with pain in the area radial tunnel, which is

–

approximately 3 to 4 cm distal to the lateral epicondyle. In contrast, pain in lateral epicondyli-

tis will localized be to the lateral epicondyle, where the ECR-B inserts (Bevelaqua et al. 2012).

RADIAL TUNNEL SYNDROME LATERAL EPICONDYLITIS (TENNIS ELBOW)

Reproduction of symptoms with:

•

Resisted extension of the third digit during elbow

extension

•

•

Resisted supination

Palpation of the radial tunnel

Reproduction of symptoms with:

•

Palpation directly on the lateral epicondyle

•

Cozen’s test: Pain on resisted wrist extension

•

Less pain on resisted supination

•

•

– – – – EDX findings:

Treatment:

NCS and needle EMG studies are typically normal

Interventions may include rehabilitation or surgical decompression.UPPER LIMB MONONEUROPATHIES

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**Lateral**

Brachialis

Brachioradialis

Biceps

Radial nerve

Deep branch

of radial nerve

(posterior

interosseous)

Superficial

branch of

radial nerve

**Medial**

Brachial

artery

Recurrent

radial artery

Arcade of

Frohse

Supinator

POSTERIOR INTEROSSEUS NERVE SYNDROME (SUPINATOR

SYNDROME, ARCADE OF FROHSE SYNDROME)

•

General:

– – This is a lesion of the PIN, which is a motor

nerve branch from the radial nerve.

•

– – – – It is considered a purely motor syndrome.

Etiology:

The nerve can be injured by compression of the

nerve at the Arcade of Frohse of the supinator

(Figure 5–99). It can also be injured by a

lipoma, ganglion cyst, ­ synovitis from RA, or a

Monteggia fracture. This is a fracture of the

proximal one-third of the ulna and ­ dislocation of

the radial head. It typically occurs from a fall on

an outstretched hand with the ­ forearm locked in

full pronation.

•

Clinical presentation:

FIGURE 5–99 The Arcade of Frohse.

– – PIN syndrome presents with painless ­ weakness

of all the PIN innervated distal extensors: EDC, EIP, ECU, EPB, EPL, APL.

– – The PIN syndrome may spare the supinator, but always spares the BR, triceps, ECR-L, ECR-

B, and anconeus, which are innervated proximally to the region of PIN entrapment.

– – A pseudo claw-hand deformity may be

­ demonstrated (finger extensor weakness).

Radial ­ deviation is noted with wrist exten-

sion (ECU weakness) and sensation is

spared.

•

EDX findings:

– NCS: Normal radial nerve SNAP.

–

Abnormal radial CMAP when recording

over PIN ­ innervated muscles.

– EMG:

–

nn Abnormal activity in the muscles

­ innervated by the PIN, which may

include the supinator. Radial nerve inner-

vated muscles proximal to the PIN lesion

are spared.

•

Treatment:

– – Interventions may include rehabilitation or

surgical release.

SUPERFICIAL RADIAL NEUROPATHY (WARTENBERG

SYNDROME) (FIGURE 5–100)

•

General: Also known as Wartenberg syndrome

or cheiralgia paresthetica. It is also known

more informally as wristwatch syndrome.

•

Etiology: Injury/compression of the superficial

FIGURE 5–100 Superficial radial neuropathy injury.

radial nerve at the wrist from a tight ­ wristwatch, tight handcuffs, peripheral IV placement, etc.

•

Clinical presentation:

– This is a pure sensory syndrome with no muscle involvement. The patient may ­ complain

–

solely of sensory abnormalities including numbness, burning, or tingling on the dorsal

radial aspect of the hand. Discomfort may be exacerbated with palmar and ulnar wrist flex-

ion or forced pronation.

•

EDX findings:

– NCS: Abnormal radial nerve SNAP but ­ normal CMAP

–

– EMG: Normal

–

•

Treatment:

– – Interventions may include removal of the compressive irritant.

Superficial

radial

nerve

Extensor

retinaculum

Extensor

carpi

radialis

Brachioradialis•

5

6

7

1

2

Musculocutaneous

nerve

Coracobrachialis

Biceps brachii

Brachialis

Lateral

cutaneous

nerve of the

forearm

Posterior

branch

Anterior

branch

Cutaneous innervation

Anterior

Posterior

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MUSCULOCUTANEOUS NERVE

Anatomy

Origin (Figure 5–101):

– – These nerve fibers originate from the C5,

C6, and C7 spinal nerve roots. These roots

continue on to contribute to the upper

trunk and lateral cord to finally form the

musculocutaneous branch.

•

Course:

ARM

•

The nerve passes along the medial aspect of

the humerus and innervates the:

– – Coracobrachialis

– – Biceps brachii

– Brachialis (also innervated by the radial nerve)

–

FOREARM

•

It continues anterior to the antecubital fossa

(lateral to the biceps tendon) to provide cutane-

ous ­ sensory innervation of the lateral forearm

as the lateral antebrachial cutaneous nerve.

Injury

MUSCULOCUTANEOUS NEUROPATHY

•

General: Injury to the distal portion of the nerve

is more common than to the proximal portion.

•

Etiology:

– – The nerve can be injured by entrapment

proximally from the coracobrachialis or

FIGURE 5–101 The musculocutaneous nerve.

distally, where it runs superficially. It can also be injured by trauma (proximal humeral frac-

tures, shoulder ­ dislocation, gunshot wounds), compression, and phlebotomy.

•

Clinical presentation:

– Involves musculocutaneous nerve inner-

–

vated muscles including biceps and brachia-

lis. The ­ coracobrachialis is typically spared.

– The patient may complain of elbow flexion

–

weakness (biceps, brachialis weakness) and

abnormal sensation over the lateral forearm.

EDX findings:

– NCS:

–

nn Abnormal SNAP in the lateral

­ antebrachial cutaneous nerve

nn Abnormal CMAP to the biceps brachii

– EMG:

–

nn Abnormal activity in the brachialis and

biceps brachii

•

Treatment:

– – Interventions may include rehabilitation or

surgical release.

AXILLARY NERVE

Anatomy

•

Origin (Figure 5–102):

FIGURE 5–102 The axillary nerve.

– – These nerve fibers originate from the C5 and C6 spinal nerve roots. They continue on to con-

tribute to the upper trunk and posterior cord to finally form the axillary nerve.

C5

Teres minor

Deltoid

5

6

7

1

2

•

Axillary nerve

Radial nerve

Upper lateral

cutaneous

nerve of arm

Cutaneous

innervation

Side view

Posterior viewUPPER LIMB MONONEUROPATHIES

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•

Course:

– – The nerve passes inferior to the ­ glenohumeral joint, through the quadrilateral space of the

axilla and to the posterior aspect of the humerus. It innervates the:

nn Teres minor

nn Deltoid

nn Upper lateral cutaneous nerve

Injury

AXILLARY NEUROPATHY

•

General:

– – The nerve runs through the quadrangular space. This is bordered by the humerus, long head

of the triceps brachii, teres minor, and teres major.

•

Etiology:

– – The nerve can be injured by traction or compression from a shoulder dislocation, humeral

head fracture, or improper axillary crutch use.

•

Clinical presentation:

– – Involves all axillary nerve innervated muscles (deltoid and teres minor)

– The patient may complain of weakness of shoulder flexion and abduction (deltoid weakness)

–

and external rotation (teres minor weakness). There may also be abnormal sensation of the lateral

shoulder.

•

EDX findings:

– NCS:

–

nn SNAP not available

nn CMAP: Abnormal

– EMG:

–

nn Abnormal activity in the axillary nerve innervated muscles (deltoid, teres minor). The

anterior and lateral heads of the deltoid show abnormalities more commonly than the

posterior head.

•

Treatment:

– – Interventions may include rehabilitation or surgical decompression, as well as discontinua-

tion of crutch use.

SUPRASCAPULAR NERVE

Anatomy

•

Origin (Figure 5–103):

– These nerve fibers originate from the C5

–

and C6 spinal nerve roots. They continue

on to contribute to the upper trunk, which

branches off to form the suprascapular nerve.

•

Course:

NECK

•

It passes the posterior triangle of the neck and

runs beneath the trapezius to the superior

margin of the scapula.

•

It runs through the suprascapular notch,

which is covered by the transverse scapu-

lar ligament and branches to innervate the

supraspinatus. The nerve then wraps around

the spinoglenoid notch to innervate the

infraspinatus.

Injury

SUPRASCAPULAR NEUROPATHY

•

General:

– – This is the only peripheral nerve injury at the trunk level. It is the most commonly involved

nerve in neuralgic amyotrophy (Figure 5–104).

C4

Dorsal scapular nerve

(n. to rhomboids)

Suprascapular

nerve

4

5

6

7

1

Supraspinatus

Infraspinatus

Rhomboid

major

Rhomboid

minor

Levator

scapulae

FIGURE 5–103 The suprascapular nerve.5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

•

C5

C6

Suprascapular

nerve

Transverse

scapular

ligament

Coracoid

process

Supraspinatus

Infraspinatus

•

•

406 •

Etiology:

– – The nerve can be injured from trauma,

including forced scapular protraction,

­ penetrating wounds, traction from a

­ massive rotator cuff tear, stinger/Erb’s

palsy; compression from spinoglenoid

ganglions, hematoma, suprascapular

or spinoglenoid notch entrapment, or

­ paralabral cyst.

– Activities involving exaggerated shoulder

–

movements, including sports with repeti-

tive overhead throwing/hitting such as

volleyball, baseball, and lacrosse, may also

injure the nerve. Volleyball more com-

monly injures branches to the IS muscle.

Clinical presentation:

– – Injury to the nerve at the suprascapular

notch results in weakness in both SS and

IS muscles. Nerve injury at the spinogle-

noid notch will result in weakness only

FIGURE 5–104 Suprascapular nerve injury sites.

in the IS muscle.

– – The patient may present with weakness

in abduction (SS) and/or external rotation (IS) of the glenohumeral joint.

EDX findings:

– NCS:

–

nn SNAP not available

nn CMAP: Abnormal

– EMG:

–

nn Abnormal activity in the IS only if entrapment is at the spinoglenoid notch or both SS and

IS muscles if nerve entrapment is at the ­ suprascapular notch.

Treatment:

– – Interventions may include rehabilitation, paralabral cyst aspiration, or surgical

decompression.

LONG THORACIC NERVE

Anatomy

•

Origin (Figure 5–105):

– – These nerve fibers originate directly

from the C5, C6 and C7 spinal nerve

roots.

•

Course:

NECK

•

The nerve runs distally along the thoracic

wall to innervate the serratus anterior

(SALT [Serratus Anterior Long Thoracic]).

Injury

SCAPULAR WINGING

•

General:

– – The long thoracic nerve innervates the

serratus anterior, which protracts and

laterally rotates the scapula.

– – The spinal accessory nerve innervates

the trapezius, which retracts, elevates,

and medially rotates the scapula.

Long thoracic nerve

Medial anterior

thoracic nerve

Lateral anterior

thoracic nerve

5

6

7

1

2

Pectoralis minor

Serratus

anterior Pectoralis major

(Sternocostal portion)

(Clavicular portion)

FIGURE 5–105 The long thoracic nerve.•

•

MEDIAL SCAPULAR WINGING LATERAL SCAPULAR WINGING

Nerve injury Long thoracic nerve Spinal accessory nerve

Affected muscle/function Serratus anterior

(Normal function: ­ protracts and laterally

rotates the scapula)

Trapezius

(Normal function: retracts and medially

rotates the scapula)

Position of scapula

(Medial border)

Scapula retracted and elevated

Medial scapular border closer to midline

due to unopposed retraction from the

trapezius

Scapula protracted and depressed

Medial scapular border is pulled away

from midline by unopposed ­ protraction

from the serratus anterior

Shoulder abduction

causes

Decreases winging Increases winging

•

•

LOWER LIMB MONONEUROPATHY

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– – The serratus anterior and trapezius help to balance scapular motion in opposing directions.

Dysfunction in one of the muscles due to nerve injury can result in scapular winging.

– – There are two main types of shoulder winging that should be differentiated from each other

(Table 5–31):

nn Medial winging is due to serratus anterior weakness from a long thoracic nerve injury.

nn Lateral winging results from trapezius weakness from a spinal accessory nerve injury.

Etiology:

– – The nerve can be injured by traction from a fall, MVA, sports activities, or shoulder bags.

Clinical presentation:

– – Shoulder pain and weakness as presenting symptoms with scapular winging.

TABLE 5–31 Scapular Winging: Medial Versus Lateral Scapular Winging

EDX findings:

– NCS:

–

nn SNAP: Not available

nn CMAP: Abnormal

– EMG:

–

nn Abnormal activity in the serratus anterior with a long thoracic nerve injury. Abnormal

activity in the trapezius and SCM indicates injury to the spinal accessory nerve.

Treatment:

– – Interventions may include rehabilitation or surgical procedure.

– Treatment of serratus anterior injury/injury to the long thoracic nerve: (a) acute stage, pain

–

reduction and ROM exercise; (b) intermediate stage, passive stretching of the rhomboids,

­ levator scapulae, and pectoralis minor; (c) late stage, strengthening exercise of all shoulder

girdle muscles, including the trapezius.

– Treatment of trapezius palsy/injury to the spinal accessory nerve: Will involve physical therapy

–

to adequately strengthen adjacent muscle groups, including rhomboids and levator scapulae.

– – Surgical repair with a dynamic muscle transfer is recommended if the patient fails conserva-

tive treatment.

n

LOWER LIMB MONONEUROPATHY

LATERAL FEMORAL CUTANEOUS NERVE

Anatomy

•

Origin (Figure 5–106):

– – These pure sensory nerve fibers originate from the L2 and L3 nerve roots from the posterior

­ divisions of the lumbar plexus.L2

2

3

4

Lateral cutaneous

nerve of thigh

Posterior

branch

Anterior

branch

Obturator

nerve

Posterior

branch

Anterior

branch

Obturator

externus

Adductors:

longus

magnus

brevis

Gracilis

Cutaneous

branch

Field of lateral

cutaneous

nerve of thigh

Cutaneous. field of

obturator nerve

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•

Course:

PELVIS/THIGH

•

The nerve continues distally to pass over the

iliacus toward the anterior iliac spine. It passes

­ underneath the inguinal ligament to provide

sensation to the lateral thigh.

INJURY

LATERAL FEMORAL CUTANEOUS NEUROPATHY

(MERALGIA PARESTHETICA)

•

General:

– This nerve injury is also known as meralgia

–

­ paresthetica and is mainly a clinical diagnosis.

•

Etiology:

– – The nerve can be injured from chronic com-

pression by a repeated low grade trauma,

from a ­ protuberant abdomen, pregnancy, or

tight clothing.

– – Diabetes, tumor, infection, and rapid weight

gain or weight loss can also affect the nerve.

– Iatrogenic cause may be present, including

–

incisions for lower abdominal/pelvic surgeries

and laparoscopic hernia repairs (Ivins, 2000).

FIGURE 5–106 The lateral femoral cutaneous nerve.

Clinical presentation:

– – This is a pure sensory syndrome with no muscle involvement.

– The patient may report sensory complaints in the lateral thigh, including pain, numbness,

–

burning, or a dull ache, exacerbated with hip extension or hyperflexion, prolonged squatting,

or driving.

EDX findings:

– NCS:

–

nn Abnormal lateral femoral cutaneous

nerve SNAP. CMAP not available.

– EMG:

–

nn Not available

Treatment:

– Interventions may include rehabilitation,

–

NSAIDs, cortisone injections, surgical release.

– – – – Symptoms may be self-limited

In addition, removal of compressive cloth-

ing should occur (e.g., wide belt, compres-

sive athletic clothing).

FEMORAL NERVE

Anatomy

•

Origin (Figure 5–107):

– – These nerve fibers originate from the L2,

L3, and L4 spinal nerve roots, continue

on as the ­ posterior division of the lumbar

plexus, and terminate as the femoral nerve.

•

Course:

– The nerve runs through the psoas muscle. It

–

travels under the inguinal ligament lateral to

the femoral artery and through the femoral

triangle to innervate the following muscles:

nn Iliacus

nn Pectineus (1/2)

•

•

•

2

3

4

Iliacus

**Quadriceps:**

Rectus

femoris n.

Vastus

lateralis n.

Vastus

medialis n.

Vastus

intermedius n.

Femoral n.

Pectineus n.

Sartorius n.

Medial cutaneous

n. of thigh

Intermediate

cutaneous n. of thigh

Saphenous n.

Infrapatellar branch

Terminal branch

**Cutaneous**

**distribution**

**from anterior**

**aspect Cutaneous**

**distribution**

**from medial**

**aspect**

FIGURE 5–107 The femoral nerve.LOWER LIMB MONONEUROPATHY

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– – nn Sartorius

nn Quadriceps muscles:

nn Rectus femoris

nn Vastus lateralis

nn Vastus intermedius

nn Vastus medialis

The saphenous nerve, a pure sensory nerve, branches from the femoral nerve and travels

through the adductor canal to the rest of the leg and provides sensation to the anteromedial

thigh, calf, and foot.

INJURY

FEMORAL NEUROPATHY

•

General:

– – This nerve is the largest branch of the lumbar plexus.

•

Etiology:

– – The nerve can be injured by trauma (e.g., fracture, abdominal/pelvic surgery, anterior total

hip arthroplasty, direct trauma from cardiac catherization), compression from a retroperito-

neal ­ hematoma, tumor, or inguinal ligament.

•

– – – – The most common cause is iatrogenic from abdominal or pelvic surgery.

Clinical presentation:

Involved muscles include all femoral nerve innervated muscles. The patient may complain

of weakness of knee extension (quadriceps), knee instability, and decreased sensation over

the anterior thigh and medial leg. Hip flexion weakness is noted with injuries occurring

above the inguinal ligament.

•

EDX findings:

– NCS:

–

nn Abnormal saphenous nerve SNAP

Abnormal CMAP to the rectus femoris

nn – EMG:

–

nn Abnormal activity in the femoral nerve innervated muscles. Involvement of obturator

nerve innervated muscles (e.g., adductor longus) and/or lumbar paraspinals could indi-

cate an L3/4 radiculopathy over a femoral neuropathy.

•

Treatment:

– – Interventions may include rehabilitation or surgical procedure.

DIABETIC AMYOTROPHY

•

General:

– – – – Diabetic amyotrophy is the most common cause of femoral neuropathy.

Diabetic amyotrophy is a proximal diabetic neuropathy distinct from other types of distal

diabetic peripheral neuropathies.

– It is also known as a lumbosacral radiculoplexus neuropathy, as it can involve the plexus and

–

nerve roots, as well as peripheral nerves. It predominantly affects the lumbosacral plexus.

•

Etiology:

– – The exact cause of diabetic amyotrophy is unknown but is believed to result from a multifo-

cal immune-mediated microvasculitis. The nerve is believed to be injured from an abnor-

mality of the vaso-nervorum due to diabetes mellitus. Nerve biopsy shows multifocal nerve

fiber loss suggesting ischemic injury and perivascular infiltrate.

•

– – – – This amyotrophy has been noted to occur after marked weight loss.

Clinical presentation (Pasnoor et al., 2013):

It typically affects an older group of diabetics, more frequently males, usually over age 50.

Most patients have type 2 diabetes mellitus.

– This neuropathy begins with severe unilateral pain in the lumbar region or proximal lower

–

extremity, which commonly spreads to the contralateral side within weeks to months. Patients

then develop weakness and atrophy of the proximal > distal lower extremity musculature.

– – – – There is typically associated weight loss that precedes symptoms.

The patient may complain of asymmetric thigh pain, knee extension weakness (quadriceps),

and atrophy. Loss of the patellar reflex may also occur.410 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

•

•

EDX findings:

– NCS:

–

nn Abnormal SNAP

nn Abnormal CMAP

– EMG:

–

nn Abnormal activity seen in femoral innervated muscles (±), adductors, iliopsoas, and

paraspinals

Treatment:

– – Diabetic amyotrophy is a self-limited condition, but the recovery process is gradual and

occurs over a period of months. There are some patients who are left with residual lower

extremity weakness.

– – Treatment is focused around pain control and optimizing glycemic control. Physical therapy

can assist in improving strength and functional mobility.

SAPHENOUS NEUROPATHY

•

General:

– – – – This nerve is the largest and longest sensory branch of the femoral nerve.

It supplies sensation to the medial aspect of the leg, the medial malleolus, and medial arch of

the foot.

•

Etiology:

– – The nerve can be entrapped in the hip adductor canal (aka subsartorial or Hunter’s canal) or

between the sartorius and gracilis.

– – It can also be related to iatrogenic trauma, including knee arthroscopy, and meniscectomy or

­ vascular surgery, such as catheterization or thrombectomy.

•

Clinical presentation:

– – – – This is a pure sensory syndrome with no muscle involvement.

The patient may complain of medial knee pain (infrapatellar branch) with abnormal sensa-

tion radiating distally along the medial aspect of the leg and foot.

•

EDX findings:

– NCS:

–

nn Abnormal saphenous nerve SNAP

Normal femoral nerve CMAP

– EMG:

–

nn Normal EMG of femoral nerve inner-

vated muscles.

•

Treatment:

– Interventions may include rehabilitation or

–

surgical procedure.

L2

2

3

4

Lateral cutaneous

nerve of thigh

OBTURATOR NERVE

Anatomy

•

Origin (Figure 5–108):

– – Nerve fibers originating from the L2, L3,

and L4 spinal nerve roots continue on as

the anterior portion of the lumbar plexus

in front of the sacroiliac joint to form the

obturator nerve, which innervates the hip

adductor musculature.

•

Course:

– – The nerve passes through the psoas major

and obturator foramen to innervate:

nn Pectineus (1/2)

nn Adductor brevis

nn Adductor longus

nn Adductor magnus

Posterior

branch

Anterior

branch

Obturator

nerve

Posterior

branch

Anterior

branch

Obturator

externus

Adductors:

longus

magnus

brevis

Gracilis

Cutaneous

branch

Field of lateral

cutaneous nerve

of thigh

Cutaneous field of

obturator nerve

FIGURE 5–108 The obturator nerve.LOWER LIMB MONONEUROPATHY

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nn Obturator externus

nn Gracilis

nn Cutaneous branch

Injury

OBTURATOR NEUROPATHY

•

General:

– – This nerve injury can occur in conjunction with a femoral nerve injury.

– – The obturator nerve gives sensation to the medial aspect of the thigh and innervates the

muscles in the medial compartment of the thigh, which includes the obturator externus,

adductor brevis, adductor longus, adductor magnus muscles, and gracilis muscles.

•

Etiology:

– – The nerve can be injured by surgery, compression from a hematoma or tumor, a pelvic frac-

ture, or obturator hernia.

•

Clinical presentation:

– – Can involve all muscles innervated by the obturator nerve.

– The most common symptom is altered sensation in the inguinal/medial thigh region, includ-

–

ing pain, numbness, and/or paresthesias.

– – Patients are also usually noted to have weakness with hip ADduction and internal rotation.

During ambulation, they present with the hip externally rotated and abnormally abducted,

which results in a wide-based, circumducting gait.

•

EDX findings:

– NCS:

–

nn None available

– EMG:

–

nn Abnormal activity in the obturator nerve innervated muscles

•

Treatment:

– – Intervention may include rehabilitation or surgical repair.

SCIATIC NERVE

Anatomy

•

Origin (Figure 5–109):

– These nerve fibers from the L4, L5, S1, S2, and S3

–

roots travel through the anterior and posterior

­ divisions of the sacral plexus and eventually form

the sciatic nerve, which travels posteriorly in the leg.

•

Course:

– The sciatic nerve exits the pelvis through

–

the greater sciatic foramen between the lesser

­ trochanter and ischial tuberosity. The sciatic nerve

proper is comprised of a tibial (medial portion of

nerve) and peroneal (fibular; lateral portion) divi-

sion. It travels as one unit up to the popliteal fossa,

where it splits into its respective divisions: the

peroneal (fibular) division and the tibial division.

– Sciatic nerve muscle innervation in the a thigh.

–

– – The peroneal (fibular) division of the sciatic

nerve innervates the short head of the biceps

femoris.

– – The tibial division of the sciatic nerve

innervates:

nn Long head of the biceps femoris

nn Semitendinosus

nn Semimembranosus

nn Adductor magnus (also innervated by the

­ obturator nerve)

Adductor magnus

Semimembranosus

Semitendinosus

Biceps femoris

(long head)

Tibial n.

Sciatic n.

Biceps femoris

(short head)

Commmon

peroneal n.

Sural n.

Medial and lateral calcaneal n.

FIGURE 5–109 The sciatic nerve.5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

Ischial

tuberosity

Gluteus

medius

Superior

gluteal n.

Inferior

gluteal n.

Piriformis

Sciatic n.

Pelvic

outlet

Hamstring

muscles

•

412 INJURY

SCIATIC NEUROPATHY

•

General:

– This is the largest nerve in the human body.

–

The peroneal portion makes up the outer

two-thirds of the nerve proper.

•

Etiology:

– The nerve can be injured by hip trauma, hip

–

replacement, injection, hematoma, pelvic frac-

ture, penetrating wounds, or a gravid uterus.

– Piriformis syndrome is an uncommon type of

–

compressive sciatic neuropathy at the pelvic

(fibular) outlet that mainly affects the peroneal

(fibular) portion of the nerve as it runs inferior

or through this muscle (Figure 5–110). Because

there is no consensus on diagnosing piriformis

syndrome, estimated ­ incidence is approxi-

mately 4% (Hicks & Varacallo, 2019).

FIGURE 5–110 Piriformis syndrome.

Clinical presentation:

– – Involves all muscles innervated by the sciatic nerve.

– The patient’s complaints depend on which portion of the nerve is more involved. It can pres-

–

ent as weakness of knee flexion (hamstring weakness) and include muscles and cutaneous

innervation of both the peroneal and tibial nerves.

– The peroneal (fibular) portion is more vulnerable to injury than the tibial because the peroneal

–

portion is more fixated in the pelvis. It has larger fascicles with less protective epineural tissue.

– – The lateral hamstring and Achilles reflexes may be abnormal.

EDX findings:

– NCS:

–

nn SNAP: Abnormal super-

ficial peroneal (fibular)

and sural sensory nerves

SNAPs

nn Abnormal tibial and pero-

neal (fibular) nerve CMAPs

– EMG:

–

nn Abnormal activity in all

sciatic innervated muscu-

lature, including the short

and long heads of the

biceps femoris.

•

Treatment:

– – Interventions may include

rehabilitation or surgical

decompression.

TIBIAL NERVE

Anatomy

•

Origin (Figure 5–111):

– These nerve fibers originate

–

from the L4, L5, S1, and S2

(S3) spinal nerve roots. They

continue on as part of the sci-

atic nerve and then branch at

the popliteal fossa to finally

form the tibial nerve.

•

Flexor

digitorum

longus

**Cutaneous Distribution**

Sciatic n.

Common

peroneal n.

Tibial n.

Plantaris

Gastrocnemius

Popliteus

Soleus

Tibialis

posterior

Flexor

hallucis

longus

Sural n.

Sural n. Tibial n.

Medial

plantar n.

(MPN) Lateral

plantar n.

(LPN)

Abductor

hallucis n.

Tibial n.

Medial

plantar n.

Lateral

plantar n.

FIGURE 5–111 The tibial nerve.LOWER LIMB MONONEUROPATHY

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•

Course:

– – At the distal one-third of the thigh, the tibial portion of the nerve runs posterior to the knee

and continues distally to innervate:

nn Plantaris

nn Medial and lateral gastrocnemius

nn Popliteus

nn Soleus

– – At the soleus, it continues on as the posterior tibial nerve and innervates:

nn Tibialis posterior (TP)

nn Flexor digitorum longus (FDL)

nn Flexor hallucis longus (FHL)

– – The nerve runs under the flexor retinaculum and divides into three branches.

nn Medial plantar nerve:

nn Abductor hallucis

nn Flexor digitorum brevis

nn Flexor hallucis brevis

nn Lumbrical

nn Sensory branch

nn Lateral plantar nerve:

nn Lumbricals

nn Abductor digiti minimi

nn Quadratus plantae

nn Interossei

nn Adductor hallucis

nn Sensory branches

nn Calcaneal nerve:

nn Sensory branch

Flexor

digitorum

longus Flexor

Tibia

Tibialis

posterior

Inferior flexor

retinaculum

hallucis

longus

Posterior

tibial a./v.

Posterior

tibial n.

Medial

calcaneal n.

Lateral

plantar n.

Medial

plantar n.

Injury

TARSAL TUNNEL SYNDROME

•

General:

– – The tibial nerve may become

entrapped in the tarsal tunnel

(Figure 5–112 and Table 5-32). This

condition is ­ relatively rare.

•

Etiology:

– – The tibial nerve can be injured by com-

pression under the flexor retinaculum

in the medial ankle.

•

Clinical presentation:

– – Involves all muscles innervated by the

tibial nerve distal to the tarsal tunnel.

The patient may complain of symp-

FIGURE 5–112 The posterior tarsal tunnel.

toms related to intrinsic foot weakness.

– – Perimalleolar pain, numbness, and

paresthesias may extend to the toes and soles. Heel sensation may be spared due to the cal-

caneal branch departing ­ proximal to the tunnel.

– It is reproduced by ankle inversion. A positive Tinel’s sign at the medial ankle may be elicited.

–

•

EDX findings:

– NCS:

–

nn nn SNAP: Abnormal plantar nerve. Normal calcaneal nerve.

CMAP: Abnormal medial and lateral plantar nerve

– EMG:

–

nn Abnormal activity in the tibial nerve innervated muscles

•

Treatment:

– – Interventions may include rehabilitation or surgical release.414 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

TABLE 5–32 Tarsal Tunnel Contents

Tom Tibialis posterior

Dick Flexor digitorum longus

And Posterior tibial artery

Very Posterior tibial vein

Nervous Tibial nerve

Harry Flexor hallucis longus

*Note:* Mnemonic for the contents of the tarsal tunnel is as follows: Tom, Dick, And Very Nervous Harry.

COMMON PERONEAL (FIBULAR) NERVE

Anatomy

•

Origin:

– – These nerve fibers originate from the L4–S2 spinal nerve roots. They continue on as the sci-

atic nerve, which divides to form the peroneal (fibular) nerve.

•

Course (Figures 5–113 and 5–114):

– – In the posterior thigh, the peroneal (fibular) fibers within the sciatic nerve innervate the

short head of the biceps femoris, the only peroneal-innervated muscle above the level of the

­ fibular neck. More distally, the sciatic nerve bifurcates above the popliteal fossa into the com-

mon ­ peroneal (fibular) and tibial nerve.

Superficial Peroneal (Fibular) Nerve innervates:

•

Peroneus longus

•

Peroneus brevis

•

Medial cutaneous nerve

•

Lateral cutaneous nerve

Deep Peroneal (Fibular) Nerve innervates:

•

Tibialis Anterior

•

Extensor Digitorum Longus

•

Extensor Hallucis Longus

•

Peroneus tertius

•

Extensor Digitorum Brevis

•

First dorsal interossei

•

Dorsal distal cutaneous nerve

Lateral cutaneous

n. of calf

Common

peroneal n.

Deep

peroneal n.

(cut)

Superficial

peroneal n.

Peroneus

longus

Peroneus

brevis

Medial

cutaneous

branch

Lateral

cutaneous

branch

**Cutaneous**

**Distribution**

Common

peroneal n.

Deep

peroneal n.

Superficial

peroneal n.

(cut)

**Anterior**

**Lateral**

**Cutaneous**

**Distribution**

Tibialis

anterior

Extensor

digitorum

longus

Extensor

hallucis

longus

Peroneus

tertius

Extensor

digitorum brevis

1st dorsal

interosseous

Dorsal digital

cutaneous n.

FIGURE 5–113 The superficial peroneal (fibular) nerve.

FIGURE 5–114 The deep peroneal (fibular) nerve.LOWER LIMB MONONEUROPATHY

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Superficial

peroneal n.

Accessory

deep

peroneal n.

Lateral

malleolus

Accessory

deep

peroneal n.

Superior

extensor

retinaculum

Inferior

extensor

retinaculum

Extensor

digitorum

brevis

FIGURE 5–115 The accessory peroneal

(fibular) nerve.

**Posterior Lateral View**

Biceps

femoris

tendon

ACCESSORY PERONEAL (FIBULAR) NERVE

•

General (Figure 5–115):

– – This is an anomalous nerve branch from the

superficial peroneal (fibular) nerve that travels

posterior to the lateral ­ malleolus to innervate some or

all of the extensor digitorum brevis (EDB).

•

– – – – Present in one-third of the population.

EDX findings:

Lower peroneal (fibular) CMAP (recording at EDB)

with stimulation at the ankle compared to stimula-

tion at the knee, with greater amplitudes above the

fibular head. The presence can be confirmed by

stimulating behind the lateral malleolus, which will

also elicit a CMAP from the EDB. With this anoma-

lous innervation, the EDB can be spared even with a

deep peroneal (fibular) nerve injury.

Injury

COMMON PERONEAL (FIBULAR) NEUROPATHY

•

General (Figure 5–116):

– – The most common site of this nerve injury is at the

fibular head.

•

Etiology:

– – The nerve can be injured by compression from

­ prolonged leg crossing, weight loss, poor ­ positioning

during surgery, poor cast application, prolonged

­ squatting position (strawberry ­ pickers’ palsy), and

metabolic disorders such as diabetes.

•

Clinical presentation:

– – The muscles involved include all muscles supplied

by the deep and superficial branches of the common

­ peroneal (fibular) nerve (short head of the biceps

­ femoris is spared).

FIGURE 5–116 Common peroneal nerve

proximity to the fibula head.

– The patient may complain of weakness of the dor-

–

siflexors (tibialis anterior [TA], extensor ­ digitorum

longus [EDL], extensor hallucis longus [EHL]), resulting in a foot drop or foot slap and a

steppage gait. Weakness of only the ankle dorsiflexors and ankle evertors helps to clini-

cally ­ differentiate a peroneal (fibular) nerve injury from an L5 radiculopathy. A radicu-

lopathy will also involve the ankle invertors.

– – Sensory loss may be noted over the distribution of the deep and superficial peroneal (fibular)

nerves.

– – A positive Tinel’s sign may be noted at the fibular head.

•

EDX findings:

– NCS:

–

nn Abnormal superficial peroneal (fibular) SNAP

nn CMAP: Abnormal waveform change from below to above the fibular head. The TA can be

used if the EDB is atrophied.

– EMG:

–

nn Abnormal activity in the muscles innervated by the superficial and deep peroneal

(fibular) nerves. The short head of the biceps femoris is spared (sciatic nerve, peroneal

(fibular) division).

•

Treatment:

– – Interventions may include rehabilitation or surgical procedure.

Gastrocnemius

Common

peroneal

Fibular head

Peroneus longus

Gastrocnemius•

Extensor

dig. longus

Extensor

hal. longus

Tibialis

anterior

•

Fibula

Inferior

extensor

retinaculum

Superior

extensor

retinaculum

Tibia

Dorsalis

pedis a. & v.

Deep

peroneal n.

•

Extensor

digitorum

brevis

Sensory area

of deep

peroneal n.

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DEEP PERONEAL (FIBULAR) NEUROPATHY

General:

– – Injury to this nerve is associated with

an anterior tarsal tunnel syndrome

(Figure 5–117), which refers to entrap-

ment of the deep peroneal (fibular)

nerve under the inferior ­ extensor reti-

naculum of the ankle.

Etiology:

– – The terminal portion of the deep

peroneal (fibular) nerve can be

injured by compression from

footwear (high boots, tight shoes, high-

heeled shoes) or intrinsic etiologies

(osteophyte, ganglion cyst, lipoma) as

it passes under the extensor retinacu-

lum as well as trauma (ankle sprains or

fractures).

Clinical presentation:

– The patient may complain of foot

–

pain, weakness, and atrophy (EDB).

There may also be ­ numbness and par-

esthesias in the first and second web

space. Pain may be located over the

­ dorsum of the foot and relieved with

motion.

EDX findings:

– NCS:

–

nn SNAP: Abnormal deep peroneal

(fibular) SNAP (Abnormal findings to

the first web space)

nn CMAP: Abnormal peroneal (fibular) CMAP with recording at the EDB (abnormal findings

to the EDB).

– EMG: Abnormal activity in deep peroneal (fibular) innervated muscles

–

Treatment:

– – Interventions may include rehabilitation or surgical resection.

SUPERFICIAL PERONEAL (FIBULAR) NEUROPATHY

General:

– – After innervating the peroneus longus and brevis, the nerve continues distally as a pure

sensory nerve.

Etiology:

– – The nerve can be injured by compression from trauma, ankle sprain, compartment syn-

drome, or a lipoma.

Clinical presentation:

– Patients complain of pain, numbness, and/or paresthesias in the loss of sensation in distal

–

­ anterolateral calf and dorsal foot (except for the first webspace). Foot eversion weakness can

be noted if the injury is proximal to the nerve innervating the peroneus longus and peroneus

brevis muscles.

EDX findings:

– NCS:

–

nn Abnormal superficial peroneal (fibular) SNAP. Normal deep peroneal (fibular) SNAP.

nn Superficial peroneal (fibular) CMAP not available. Normal deep peroneal (fibular)

CMAP.

•

FIGURE 5–117 The anterior tarsal tunnel.

•

•

•

•

•LOWER LIMB MONONEUROPATHY

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Sciatic n.

Tibial n.

Medial sural

cutaneous n.

Common

peroneal n.

Lateral sural

cutaneous n.

Sural n.

•

– EMG:

–

nn Treatment:

Abnormal activity in the peroneus longus and/or peroneus brevis but normal findings in

deep peroneal (fibular) nerve innervated muscles.

– – Interventions may include rehabilitation or surgical release.

SURAL NERVE

Anatomy

•

Origin (Figure 5–118):

– These nerve fibers originate from branches of the

–

tibial and common peroneal (fibular) nerves.

•

Course:

– – The sural nerve travels from the proximal poste-

rior calf to the lateral malleolus of the proximal

ankle. It supplies cutaneous innervation to the

lateral calf and foot.

INJURY

SURAL NEUROPATHY

•

General:

– – This is a pure sensory nerve.

•

Etiology:

– – The nerve can be injured by compression from

tight socks, a Baker’s cyst or ganglion cyst, or

trauma (e.g., laceration) (Bryan et al., 1999).

•

Clinical presentation:

– – This is a pure sensory nerve with no muscle

involvement. The patient may complain of

abnormal sensations to the lateral calf and foot.

A positive Tinel’s sign may be elicited along the

course of the nerve.

•

EDX findings:

– NCS:

–

nn Abnormal SNAP. CMAP not applicable

nn Note: Decreased or absent sural responses can be seen as a normal finding in elderly

patients (Tavee et al., 2013).

– EMG: Not applicable

–

•

Treatment:

– – Interventions may include rehabilitation or surgical procedure.

SUPERIOR AND INFERIOR GLUTEAL NERVES

Anatomy

SUPERIOR GLUTEAL NERVE

•

Origin:

– – These nerve fibers originate from the L4, L5, S1 spinal nerve roots.

•

Course:

– – In the pelvic region, the nerve passes through the sciatic notch superior to the piriformis

muscle to innervate:

nn Gluteus medius

nn Gluteus minimus

nn Tensor fascia lata (TFL)

FIGURE 5–118 The sural nerve.•

•

•

•

•

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INFERIOR GLUTEAL NERVE

Origin:

– – – – Course:

These nerve fibers originate from the L5–S2 spinal nerve roots.

In the pelvic region the nerve supplies the gluteus maximus.

Injury

•

General:

– – These nerves are typically injured by iatrogenic causes, such as hip joint replacement,

improper intramuscular injection, or pelvic masses.

Clinical presentation:

– A superior gluteal neuropathy presents with weakness of hip abduction and external rotation.

–

A Trendelenburg gait demonstrating a pelvic tilt to the strong side will be seen.

– – In contrast, an inferior gluteal neuropathy presents with weakness of hip extension. Sensation

is spared.

EDX findings:

– NCS:

–

nn No SNAP or CMAP studies exist

– EMG:

–

nn Superior gluteal neuropathy: Abnormal findings in the gluteus medius, gluteus minimus,

and TFL only

nn Inferior gluteal neuropathy: Abnormal findings in the gluteus maximus only

MONONEURITIS MULTIPLEX

General:

– – – – Also known as mononeuropathy multiplex or multifocal neuropathy.

This is a type of multifocal peripheral neuropathy in which nerve damage occurs in two or

more different nerve areas.

– – The condition refers to a group of symptoms (syndrome) rather than a disease. It can encom-

pass a constellation of symptoms that includes numbness, paresthesias, focal weakness or

paralysis, and bowel/bladder dysfunction.

Etiology:

– – Axonal injury can be caused by multiple etiologies, including inflammation (vasculitis),

vascular compromise (occlusion), compression, and infection.

– – It is associated with a number of conditions:

nn Rheumatologic (polyarteritis nodosa, RA, systemic lupus erythematosus (SLE), sclero-

derma, Wegener’s granulomatosis, Sjrögren’s)

nn Diabetes mellitus—most common; amyloidosis

nn Demyelinating (acute inflammatory demyelinating polyneuropathy [AIPD], chronic

inflammatory demyelinating polyneuropathy [CIPD])

nn Infections (Lyme, leprosy, HIV, hepatitis A, B, C)

nn Cancer (neurofibroma, malignant invasion)

Clinical presentation:

Based on etiology and which peripheral nerves, cranial nerves, nerve roots, or plexus loca-

tions are involved.

Onset can be acute, subacute, or of a gradual presentation and pathology is asymmetric.

Sensation becomes abnormal and primarily affects pain and temperature (small fiber nerves)

in a poorly localized pattern along with migratory arthralgias and myalgias.

Foot drop or give-way weakness can develop in an asymmetric distribution and muscle

stretch response (MSR) will be absent.

Abnormal findings on studies are based on etiology, but nerve infarctions cause primarily

axonal degeneration and less commonly segmental demyelination. This will affect recovery,

with axonal involvement being more prolonged and incomplete.

– – Nerve biopsy may provide definitive diagnosis.

EDX findings:

– EDX studies will demonstrate a typical pattern but are dependent on which nerve is involved.

–

•

•

– – – – – – – – – – •PERIPHERAL POLYNEUROPATHY (PERIPHERAL NEUROPATHY)

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EDX Findings in Mononeuritis Multiplex

MINIMAL DEMYELINATION FEATURES AXONAL DEGENERATION

EDX Findings:

Normal CV

Increased in chronic conditions

Normal temporal dispersion

No conduction block

EDX Findings:

Decreased amplitudes

EMG abnormalities of spontaneous activity with decreased

recruitment and polyphasic potentials of increased

­ duration and amplitude, demonstrating collateral sprouting.

CV, conduction velocity; EDX, electrodiagnostic; EMG, electromyography.

n

PERIPHERAL POLYNEUROPATHY

(PERIPHERAL NEUROPATHY)

•

•

•

•

•

Peripheral neuropathies (polyneuropathies) have numerous causes and are typically catego-

rized as either inherited or acquired in origin.

They can present with acute clinical symptoms or demonstrate a more insidious progression.

EDX findings can give further insight on not only the type of neuropathy but the nerve fibers

affected to provide a greater understanding of the disorder.

Peripheral polyneuropathies can affect the myelin and/or axons of the peripheral nerves. If

­ demyelination becomes extensive, it can be accompanied by mild axonal damage.

Determining a primary and predominating process is helpful in understanding the course of the

­ disorder. The primary process can help determine the etiology of a neuropathy, while the predom-

inant process can help assess the prognosis. They can be described as diffuse or multifocal.

PATTERN PRESENTATION

Diffuse Essentially involves all nerves in a length-dependent fashion to a relatively equal extent

Multifocal Involves one or multiple nerves in an asymmetric or patchy distribution

ETIOLOGY (TABLES 5–33 AND 5–34)

•

•

Symptom onset and location, presentation patterns, family history, and comorbidities are

important to investigate to determine whether the condition is inherited or acquired.

Certain clinical and EDX criteria are used to specifically classify the type of polyneuropathy , out-

lined on:

– – – – Table 5–33: Classification I

Table 5–34: Classification II

GENERAL OVERVIEW

Inherited Versus Acquired Peripheral Neuropathies

INHERITED PERIPHERAL NEUROPATHIES

•

This is the largest group of progressive disorders that affect the PNS. Genetic testing plays a

great role in confirming the diagnosis. These are typically categorized as:

– – Hereditary motor and sensory neuropathies (HMSN)

– – Hereditary sensory and autonomic neuropathies (HSAN)

– – Hereditary motor neuropathies (HMN)

•

Classification of inherited polyneuropathies:

– – Primary classification is based on the presentation (HMSN, HSAN, HMN)

– Subclassification is based on inheritance pattern and EDX findings (demyelinating vs. axonal)

–

– – Specific gene mutation is also relevant420 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

•

•

•

HMSN, also known as Charcot–Marie–Tooth disease, is the most commonly inherited

­ peripheral polyneuropathy and is due to a duplication mutation of the PMP-22 gene.

– – A mutation related to the PMP-22 gene is also implicated as hereditary neuropathy with

liability to pressure palsy (HNPP). This is due to a reciprocal deletion of the gene, causing

this unique diagnosis, which presents with recurrent, short-term episodes of motor and

sensory mononeuropathies. They can last from hours to a few weeks and are typically noted

in regions of entrapments or from mild trauma.

HSAN disorders are rare and primarily affect sensory and autonomic functions. There are

several subclassifications with distinguishing features causing sensory loss including pain and

temperature along with thermal dysregulation, bladder dysfunction, and cognitive deficits.

HMN is characterized by progressive atrophy and weakness of the upper and lower extremi-

ties. This arises from abnormalities in the anterior horn cells and causes atrophy of the distal

spinal muscles. They can progress to vocal cord and facial muscle paralysis.

ACQUIRED NEUROPATHIES

•

These are associated with progressive medical conditions, inflammatory processes, or iatro-

genic ­ influences, most commonly related to:

– – Diabetes mellitus (most common)

– – Acute inflammatory demyelinating polyradiculopathy (AIDP)

– – Medications

•

Acquired polyneuropathies are what we identify with most commonly due to its typical distal

onset of symptoms.

– – This is seen because of the length-dependent manner in which it affects the axons of the

nerve: It affects the distal-most part of the nerve and progresses proximally, as seen with dia-

betes mellitus, thyroid disorders, alcohol abuse, vitamin B12 deficiency, and critical illness

neuropathy.

•

Demyelinating inflammatory neuropathies can manifest as a polyradiculopathy, involving the

peripheral nerves, the nerve roots, and the cranial nerves.

•

Multiple nerves can also be affected in an asymmetric pattern manifesting either simultaneous

neuropathies or randomly throughout the body or limb.

– – These findings may be seen in ischemic processes such as vasculitic neuropathies.

•

Though less common, the upper limbs may demonstrate abnormalities first.

– This can occur in vasculitic neuropathies, chronic inflammatory demyelinating

–

polyradiculopathy (CIDP), HNPP toxicity, porphyria, inflammatory neuropathies,

diabetes mellitus, etc.

CLINICAL PRESENTATION OF POLYNEUROPATHIES

•

A classic triad has been described presenting more in the lower limbs than the upper limbs:

– – Sensory changes in a stocking/glove distribution

– – Distal weakness

– – Diminished/absent MSR

•

However, inherited polyneuropathies typically present with:

– – Sensory loss

– – Ataxia

– – Increased incidence of muscle cramping.

•

The acquired polyneuropathies tend to demonstrate:

– – Burning

– – Pain

– – Paresthesias

ELECTRODIAGNOSTIC FINDINGS

The diagnostic criteria for peripheral neuropathy consists of evaluating at least three limbs.

Abnormalities of SNAPs, CMAPs, and MUAPs are dependent on the type and timing of the pathol-

ogy affecting a nerve.PERIPHERAL POLYNEUROPATHY (PERIPHERAL NEUROPATHY)

Basic EDX Findings in Peripheral Neuropathy

•

Classic findings for a demyelinating versus axonal process are listed in the following chart.

Conditions such as ­ diabetes mellitus and uremia can present as a mixed pattern.

•

Understanding the different findings seen in a hereditary versus acquired neuropathy aids in

­ monitoring the diagnosis and can guide treatment options and prognosis.

•

EDX abnormalities can include:

– – – – – – – – – – Prolonged latencies and increased conduction velocities

Reduced or absent amplitudes

Prolonged or absent F-waves

Decreased persistence (“missing” waveforms on repetitive stimulations)

Increased chronodispersion (large latency difference between the fastest and slowest wave-

form on repetitive stimulation)

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DEMYELINATING INJURY AXONAL INJURY

NCS Prolonged distal latency

Slowed conduction velocity

Conduction block

Increased temporal dispersion

Near-normal distal latency

Near-normal conduction velocity

Reduced SNAP/CMAP amplitude

EMG No fibrillation potentials or positive sharp waves

Myokymic discharge

Decreased recruitment

(+) Fibrillation potentials

(+) Positive sharp waves

Decreased recruitment

Increased duration and amplitude

Polyphasic potentials

CMAP, compound muscle action potential; EMG, electromyography; NCS, nerve conduction studies; SNAP, sensory

nerve action potential.

NCS FINDINGS ACQUIRED NEUROPATHY HEREDITARY NEUROPATHY

Conduction block Positive Negative

Conduction velocity Focal slowing Diffuse slowing

Temporal dispersion Increased Normal

NCS, nerve conduction studies.

SPECIAL STUDIES

•

Small nerve fiber abnormalities and associated autonomic dysfunction may not be seen with

­ conventional NCS and can require additional tests. These may be considered in patients with

­ systemic symptoms such as orthostatic blood pressure changes, dry scaly skin, dry eyes, dry

mouth, etc.

•

Furthermore, small fiber neuropathies manifest as burning pain in the extremities with abnor-

mal pinprick sensation but demonstrate normal standard NCS.

Autonomic Nerve Studies

•

Norepinephrine synthesis and release:

– – Norepinephrine is the primary neurotransmitter for postganglionic sympathetic adrenergic

nerves. It is synthesized inside the nerve axon, stored within vesicles, then released by the

nerve when an AP travels down the nerve. Following are the details for release and synthesis

of norepinephrine:

nn The amino acid tyrosine is transported into the sympathetic nerve axon.

nn Tyrosine is converted to dihydroxyphenylalanine (DOPA) by tyrosine hydroxylase.

nn DOPA is converted to dopamine by DOPA decarboxylase.422 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

nn Dopamine is transported into vesicles then converted to norepinephrine by dopamine

b-hydroxylase.

nn An AP traveling down the axon depolarizes the membrane and causes calcium to enter

the axon.

nn Increased intracellular calcium causes the vesicles to migrate to the axonal membrane and

fuse with the membrane, which permits the norepinephrine to diffuse out of the vesicle

into the extracellular space.

nn The norepinephrine binds to the postjunctional receptor and stimulates the effector organ

response.

•

Sympathetic skin response:

– – The sympathetic skin response is a means of evaluating the unmyelinated, sympathetic

nerve fibers of the PNS.

– – For median nerve testing using standard electrodes, E-1 can be placed on the palm and E-2

on the dorsum of the hand. The median nerve is stimulated at the wrist and elbow at the

usual locations. Stimulation occurs over several minutes, and irregular stimulus intervals are

required to prevent nerve habituation.

– – The stimulus sources used are electrical, coughing, noises, breathing, or tactile.

nn Current: 10 to 20 mA with a pulse width of 0.1 msec

nn Sweep speed: 500 msec/cm

nn LFF: 0.5 Hz

nn HFF: 2,000 Hz

nn Upper extremity CV: 1.57 ± 0.11 m/sec

nn Lower extremity CV: 1.02 ± 0.07 m/sec

•

Sinus arrhythmia:

– – This cardiovagal innervation study is dependent on the normal heart rate variations that

occur with respiration via parasympathetic activity.

– – – – Loss of this sinus arrhythmia represents a denervation process.

The test consists of measuring the R-R ratio with an EKG machine attached to the amplifier

of the EMG.

•

Valsalva ratio:

– – During a Valsalva maneuver, the heart rate varies in response to changes in blood pressure

and intrathoracic pressure via sympathetic and parasympathetic activity. Four phases are

measured using a standard EMG machine. The findings measured in Phase 2 and Phase

4 are used for monitoring. Phase 2 should demonstrate a heart rate increase, while Phase

4 should demonstrate a decrease in the normal population.

•

Anal sphincter activity:

– – EMG recording of the external anal sphincter has continuous activity at rest. There is a brief

contraction in response to rapid rectal distension, and a preserved or increased activity dur-

ing a prolonged substantial rectal distension during defecation in healthy adults.

•

Additional studies:

– – Other studies can support the EDX findings when evaluating for peripheral neuropathies.

These can include:

nn Nerve biopsies (inflammatory neuropathy, mononeuropathy multiplex, amyloidosis)

nn Skin biopsies (small fiber neuropathy)

nn Laboratory studies (blood glucose, B12 levels, serum protein immunofixation electrophoresis)

nn Autonomic testing

The following tables outline pertinent peripheral neuropatterns as they are defined by criteria in

Classification I. Please refer to Table 5–33 as an overview for Tables 5–35 to 5–42.

DIFFERENTIAL DIAGNOSIS OF FOOT DROP

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Diffuse polyneuropathy: Diabetes

Mononeuropathy: Common peroneal (fibular); peroneal (fibular) portion of the sciatic nerve

Plexopathy

Radiculopathy: L4–L5

Central: Tumor, cerebrovascular accident (CVA), arteriovenous malformation (AVM), spinal

cord injury (SCI)TABLE 5–33 Classification I

**PERIPHERAL NEUROPATHY**

DEMYELINATION DEMYELINATION/

AXONAL LOSS AXONAL LOSS

Motor

Sensorimotor

(Uniform)

Motor > Sensory

(Segmental)

Sensorimotor Sensorimotor

Motor > Sensory

Sensory

• Multifocal motor

neuropathy

(Genetic Disorders)

• HMSN-I

• HMSN-III

• HMSN-IV

• Leukodystrophy

(Acquired Disorders)

• AIDP

• CIDP

• Arsenic

• Toxins

• Monoclonal

gammopathy

• Diphtheria

• AIDS

• Leprosy

• Lyme disease

• Diabetes

mellitus

• Uremia

• Porphyria

• Vincristine

• Lead

• AIDP

• Dapsone

• HMSN-II

• Cis-platinum

• Friedreich’s ataxia

• HSN

• Sjögren’s

syndrome

• Pyridoxine

• Crohn’s disease

• Amyloidosis

• ETOH

• Vitamin B12

• Folate

• Toxins

• Gold

• Mercury

• Paraneoplastic

syndrome

• Sarcoidosis

• Lyme disease

• HIV related

AIDP, acute inflammatory demyelinating polyneuropathy; CIDP, chronic inflammatory demyelinating polyneuropathy; HMSN, hereditary motor and sensory neuropathies; HSN, hereditary

sensory neuropathy.

PERIPHERAL POLYNEUROPATHY (PERIPHERAL NEUROPATHY)

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TABLE 5–34 Classification II

Diffuse axonal polyneuropathy Toxins—Heavy metals; Drugs—Vincristine, alcohol

Deficiency—Vitamin B6 deficiency

Metabolic—Uremia, diabetes; paraneoplastic syndrome

Hereditary—HMSN II; Infectious—Lyme’s disease, HIV

Multifocal axonal neuropathy Microangiopathic—Vasculitis, diabetes; amyloidosis; paraneo-

plastic syndrome Infectious—CMV

Metabolic—Porphyria; compression

Diffuse demyelinating polyneuropathy Hereditary—HMSN-I, IV

Deficiency—Hypothyroidism

Toxic—Amiodarone, arsenic

Multifocal demyelinating neuropathy Autoimmune—AIDP, CIDP; multiple compressions; leprosy

AIDP, acute inflammatory demyelinating polyneuropathy; CMV, cytomegalovirus; CIDP, chronic inflammatory

­ demyelinating polyneuropathy; HMSN, hereditary motor and sensory neuropathies.

TABLE 5–35 Uniform Demyelinating Mixed Sensorimotor Neuropathies: Common Disorders

DISEASE HMSN I: CHARCOT-MARIE-TOOTH

HMSN III: DÉJÉRINE

SOTTAS

HMSN IV: REFSUM’S

DISEASE

Etiology Autosomal dominant Autosomal recessive Autosomal recessive

Onset Early childhood in first 2 years Birth–infancy Approximate third decade

Clinical

presentation

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•

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•

•

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•

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•

Slowly progressive distal motor more than

sensory abnormalities

Sensory loss in the lower limbs > the upper limbs

Abnormal vibration and proprioception

Stocking/Glove pattern

Distal > Proximal weakness

Abnormal MSR

Predominantly affects the intrinsic foot and

lower leg ­ anterior ­ compartment ­ musculature:

Pes cavus and hammer toes

Bilateral foot drop: Steppage gait

Stork leg/champagne bottle leg appearance

Hypertrophy of ­ peripheral nerves (greater

auricular nerve)

Roussy–Levy syndrome: CMT associated with

an essential tremor

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Severe progression

Sensory loss

Weakness

Abnormal MSR

Hypotonic/floppy baby

Delayed milestones

Ataxia

Pes cavus

Kyphoscoliosis

Nystagmus

Deafness

•

•

•

•

•

•

•

•

•

•

Weakness

Abnormal MSR

Lower extremity

wasting

Steppage gait

Ataxia

Retinitis pigmentosa

(night blindness)

Cerebellar dysfunction

Deafness

Cardiac abnormalities

Cataract

Labs CSF: Increased protein

N Bx: Onion bulb ­ formation from focal demyelin-

ation, then remyelination

CSF: Increased protein CSF: Increased protein

N Bx: Onion bulb formation

Blood: High phytanic acid

EDX findings NCS

•

SNAP: Abnormal

•

CMAP: Abnormal, CV decreased 70%. No

temporal dispersion or conduction block

EMG: Normal

NCS

•

SNAP: Abnormal

•

CMAP: Abnormal, CV

is <6 m/sec, latency

is 7× slower than

normal

EMG: Normal

NCS

•

SNAP: Abnormal

•

CMAP: Abnormal, CV

is 10 m/sec

EMG: Normal

Treatment Rehabilitation; orthotics Rehabilitation Rehabilitation; phytanic

acid absent diet

CMAP, compound motor action potential; CMT, Charcot–Marie–Tooth; CSF, cerebrospinal fluid; CV, conduction

velocity; EMG, electromyography; HMSN, hereditary motor sensory neuropathy; MSR, muscle stretch response; N

Bx, nerve biopsy; SNAP, sensory nerve action potential.TABLE 5–36 Segmental Demyelinating Motor and Sensory Neuropathies: Common Disorders

DISEASE AIDP, GBS CIDP LEPROSY (HANSEN’S DISEASE)

Etiology Possible viral attack on the myelin and Schwann cells Possible immune mediated response Mycobacterium Leprae

Onset 1–4 weeks post illness, vaccination, or surgery Any age, peaks at 50–60 years of age Immune status dependent

Clinical presentation • Male > female

•

Ascending ­ sensory abnormalities (­ ascending numbness is

often first sign)

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•

Ascending symmetric weakness

Abnormal MSR

Possible respiratory and autonomic failure

Possibly bedridden within two days

CN involvement (Most common: CN VII affected, CN I and

II unaffected)

•

Variants: Miller-Fisher syndrome, pure sensory

•

•

•

•

•

Relapsing and remitting course

Sensory abnormalities

Symmetric ­ weakness: ­ proximal >

distal

Abnormal MSR

Less ­ cranial nerve involvement

•

•

•

•

•

Most common world-wide

neuropathy

Sensory abnormalities

Wrist drop

Foot drop

Facial palsy

Labs CSF: Increased protein, few mononuclear cells CSF: Increased protein N Bx: Foamy histiocyte invasion

EDX findings NCS

•

SNAP: Abnormal

•

CMAP: Abnormal, temporal dispersion and conduction block:

F-wave: Abnormal—first EDX sign

EMG: Normal

Poorer prognosis if:

•

CMAP: Amplitude <20% of normal, NCV <40% of normal

•

F-wave: Absent

EMG: Abnormal activity (axonal involvement)

NCS

•

SNAP: Abnormal

•

CMAP: Abnormal; increased TD

•

F-wave: Abnormal

EMG: Abnormal, if severe

NCS

•

SNAP: Abnormal

•

CMAP: Abnormal

EMG: Abnormal, if severe

Treatment Rehabilitation. Plasmapheresis, IV immunoglobulins

Steroids are ineffective; respiratory support Majority of patients

have near complete recovery with only mild permanent sequelae

within 3–6 months

Rehabilitation IV Ig, Plasmapheresis

High-dose steroids

Rehabilitation Antileprosy treatment

AIDP, acute inflammatory demyelinating polyneuropathy; CIDP, chronic inflammatory demyelinating polyneuropathy; CMAP, compound motor action potential; CMT, Charcot–Marie–

Tooth; CSF, cerebrospinal fluid; EDX, electrodiagnostic; EMG, electromyography; GBS, Guillain-Barré syndrome; IV, intravenous; MSR, muscle stretch response; N Bx, nerve biopsy,

NCS, nerve conduction study; NCV, nerve conduction velocity; SNAP, sensory nerve action potential; TD, temporal dispersion.

PERIPHERAL POLYNEUROPATHY (PERIPHERAL NEUROPATHY)

425TABLE 5–37 Axonal Motor > Sensory Neuropathies: Common Disorders

DISEASE PORPHYRIA TOXINS AIDP AXONAL HMSN II CMT–II

Etiology Defect heme synthesis Lead Vincristine

Chemotherapy

Dapsone

Leprosy treatment

Same as demyelination Autosomal dominant

Clinical

presentation

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Female > male

Lower limb pain

Limb weakness

Back and abdominal

pain

Seizures

Mental ­ status changes

Reaction to ­ medication,

e.g., barbiturates,

sulfonamides

•

•

•

•

•

•

•

Progressive onset

of upper-limb

weakness

Radial ­ neuropathy:

Wrist drop

(adult, child)

Encepha­ lopathy

(child)

Abdominal

discomfort

Blue lines in the

gums

Blindness

Epilepsy

•

•

•

Lower limb

paresthesias

Lower limb

weakness

Abnormal MSR

•

Ascending foot and

hand neuropathy

Side effects include:

Methemo­ globinemia

•

•

•

•

Areflexia

Autonomic and cranial

nerve involvement

Poorer prognosis

than with pure

demyelination

Associated with CMV

and C. jejuni infection

•

•

•

•

•

•

Onset ­ commonly in the

­ second decade

Weakness

Abnormal MSR

Less foot intrinsic

involvement

Tremor

Ataxia

Labs Urine: Deep red Blood/urine: Lead

Basophilic stippling in

RBCs, x-ray lead lines

CSF: Increased protein N Bx: No onion bulb

formation

Edx findings NCS

•

SNAP: Abnormal

CMAP: Abnormal

•

EMG

•

Abnormal

NCS

•

•

SNAP: Normal

CMAP: Abnormal

EMG

•

Abnormal radial

muscles

NCS

•

•

EMG

•

SNAP: Abnormal

CMAP: Abnormal

Abnormal

NCS

•

•

EMG

•

SNAP: Normal

CMAP: Abnormal

Abnormal

NCS

•

•

EMG

•

SNAP: Abnormal

CMAP: Abnormal

Abnormal

NCS

•

SNAP: Abnormal

•

CMAP: Abnormal with

­ preserved CV

EMG

•

Abnormal paraspinal

muscles

Treatment Rehabilitation Rehabilitation:

Penicillamine, EDTA

Rehabilitation Rehabilitation Rehabilitation Rehabilitation

AIDP, acute inflammatory demyelinating polyneuropathy; CMAP, compound muscle action potential; CMT, Charcot-Marie-Tooth; CMV, cytomegalovirus; CSF, cerebrospinal fluid;

CV, conduction velocity; EDTA, ethylenediaminetetraacetic acid; EDX, electrodiagnostic; EMG, electromyography; HMSN, hereditary motor sensory neuropathy; MSR, muscle

stretch response; NCS, nerve conduction study; N Bx, nerve biopsy; RBCs, red blood cells; SNAP, sensory nerve action potential.

426 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGYTABLE 5–38 Axonal Sensory Neuropathies: Common Disorders

DISEASE TOXINS FRIEDREICH’S ATAXIA SJÖGREN’S SYNDROME TOXINS

Etiology Cis-platinum Autosomal recessive Autoimmune disorder Pyridoxine (B6)

Clinical

presentation

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Painful paresthesias in the

hands and feet

Abnormal sensation

Side effects:

− Nephrotoxicity

−

− Ototoxicity

−

− Myelosuppression

−

− GI complaints

−

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•

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Onset: 2–16 years old

Abnormal sensation

Weakness

Abnormal MSR

Ataxia: Limb and trunk

Optic atrophy

Kyphoscoliosis

Dysarthria

Pes cavus deformity

Cardiomyopathy

Wheelchair use by 16 years

of age

•

•

•

•

Dry eyes

Dry mouth

Keratoconjunctivitis associ-

ated with rheumatoid

arthritis

Gland involvement:

− Parotid

−

− Lacrimal

−

− Salivary

−

•

•

•

•

•

Abnormal sensation

Gait disturbances

Positive Lhermitte’s sign

This may occur with doses of B6 >600 mg/day

Symptoms improve with drug withdrawal

Lab N Bx: Abnormal large axons N Bx: Abnormal large axons N Bx: Abnormal large axons N Bx: Abnormal large and small axons

EDX findings NCS

•

SNAP: Abnormal

CMAP: Normal

•

EMG

•

Normal

NCS

•

•

SNAP: Abnormal

CMAP: Normal

EMG

•

Abnormal activity (motor unit

remodeling)

NCS

•

SNAP: Abnormal

•

CMAP: Normal (can be

abnormal)

EMG

•

Abnormal (muscle

remodeling)

NCS

•

•

EMG

•

SNAP: Abnormal

CMAP: Normal

Abnormal: occ. fibs and pos. sharp waves

Treatment Drug cessation Rehabilitation Rehabilitation Stop vitamin B6

CMAP, compound motor action potential; EMG, electromyography; EDX, electrodiagnostic; GI, gastrointestinal; MSR, muscle stretch response; N Bx,nerve biopsy; NCS, nerve

conduction studies; SNAP, sensory nerve action potential.

PERIPHERAL POLYNEUROPATHY (PERIPHERAL NEUROPATHY)

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TABLE 5–39 Axonal Sensorimotor Neuropathies: Common Disorders

DISEASE ETOH AMYLOIDOSIS SARCOIDOSIS

Etiology Malnutrition or direct nerve injury Amyloid deposition in DRG Granulomatous disorder

Clinical

Presentation

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Sensory abnormalities

Foot or wrist drop

Muscle spasms

Korsakoff’s psychosis

Wernicke’s encephalopathy

± associated with a myopathy

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Sensory abnormalities

Weight loss

Ankle edema

Hepatomegaly

Purpura

Nephrotic syndrome

Congestive heart failure

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•

Low birth weight

Fatigue

Bilateral hilar

adenopathy

Uveitis

Cranial nerve

­ involvement (CN VII

most common)

Labs N Bx: Wallerian degeneration Tissue Bx: (+) ­ birefringence

with Congo red staining

Blood: Increased ESR

N Bx: Sarcoid tubercles

EDX Findings NCS

•

•

EMG

•

SNAP: Abnormal

CMAP: Abnormal

Abnormal activity

NCS

•

•

EMG

•

SNAP: Abnormal

CMAP: Abnormal

Abnormal activity

NCS

•

•

EMG

•

SNAP: Abnormal

CMAP: Abnormal

Abnormal activity

Treatment Vitamins, diet, stop alcohol con-

sumption, orthotics

Rehabilitation Rehabilitation

CMAP, compound motor action potential; CN, cranial nerve; DRG, dorsal root ganglion; EMG, electromyography;

EDX, electrodiagnostic; ESR, erythrocyte sedimentation rate; ETOH, ethyl alcohol; N Bx, nerve biopsy; NCS, nerve

conduction study; SNAP, sensory nerve action potential.

TABLE 5–40 Mixed Axonal and Demyelinating Neuropathies: Common Disorders

DISEASE DIABETES MELLITUS UREMIA

Clinical Presentation •

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•

Sensory abnormalities

Variants: Polyneuropathy,

­ mononeuropathy, autonomic

­ disorders, or amyotrophy

Most common peripheral

neuropathy in North America

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Occurs in 60% of patients with

renal failure

Sensory abnormalities

Hypersensitivity to touch

Associated with restless leg

syndrome

Labs Blood: Elevated glucose

N Bx: Small and large fiber

abnormalities

Blood: Increased nitrogen and urea

N Bx: Paranodal demyelination,

axon loss

EDX Findings NCS

•

•

EMG

•

SNAP: Abnormal

CMAP: Abnormal

Abnormal activity

NCS

•

•

EMG

•

SNAP: Abnormal

CMAP: Abnormal

Abnormal activity

Treatment Rehabilitation: Control blood sugar Rehabilitation: Dialysis, kidney

transplant

CMAP, compound motor action potential; EDX, electrodiagnostic; EMG, electromyography; N Bx, nerve biopsy;

NCS, nerve conduction studies; SNAP, sensory nerve action potential.PERIPHERAL POLYNEUROPATHY (PERIPHERAL NEUROPATHY)

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TABLE 5–41 Demyelinating Motor Neuropathy

DISEASE MULTIFOCAL MOTOR NEUROPATHY (MMN)

Etiology Immune-mediated disorder causing inflammatory demyelination and remyelination

Clinical

Presentation

Slowly progressing focal weakness

Spreading fasciculations and cramps

Atrophy and myokymia

Asymmetric reduced MSR

Sensation is normal

Resembles MND

Labs Nerve Bx: Endoneurial edema, lymphocytic inflammation, reduced myelin density, onion bulb

formation (Findings resemble CIPD, except MMN only affects motor nerves.)

Blood: Increased anti-GM1 antibody titers

EDX

Findings

•

•

•

•

•

SNAP: Typically normal, though mild changes have been noted

CMAP: Latencies typically abnormal, amplitudes can be normal in weak muscles or show an

80% drop, decreased CV

MMN is defined by multifocal motor conduction block. More than one site of CB can occur in a

single motor nerve.

F-wave: Abnormal

EMG: Abnormal spontaneous activity, including fasciculations and myokymic discharges

Other Findings that help distinguish MMN from MND:

•

•

•

•

In MMN activity is confined to the muscles of clinical weakness

In MND it is diffusely distributed

In MMN activity can be traced back to peripheral nerve territories

In MND it can be traced to a spinal segmental pattern

Treatment High dose IV-Ig

CB, conduction block; CIPD, chronic inflammatory demyelinating polyneuropathy; CMAP, compound motor action

­ potential; CV, conduction velocity; EDX, electrodiagnostic; IV, intravenous; MND, motor neuron disease; MMN, mul-

tifocal motor neuropathy; MSR, muscle stretch response; SNAP, sensory nerve action potential.

TABLE 5–42 HIV-Related Neuropathies

FIVE MAJOR CATEGORIES

1. Distal Symmetric Polyneuropathy: This is the most common type of neuropathy. It primarily affects sensory

and autonomic fibers, with motor disruption occurring in advanced cases. Painful paresthesias begin in the toes

followed by the fingers and advance proximally up the extremities.

2. Inflammatory Demyelinating Polyneuropathy: This presents in a similar manner as AIDP or CIDP. However, pleocy-

tosis in the CSF with elevated protein distinguishes it from idiopathic AIDP/CIDP.

3. Mononeuropathy Multiplex: Thrombosis of the vasa nervorum leads to multiple lesions in various nerves. This

causes primarily axonal loss with relative myelin sparing. This results in abnormal spontaneous activity on needle

exam, but normal NCS with decreased amplitudes.

4. Progressive Polyradiculopathy: This results from cytomegalovirus causing severe asymmetrical pain, numbness,

and motor deficits in the legs. Bowel and bladder dysfunctions along with impaired MSR are also noted.

5. Autonomic Neuropathy: A group of symptoms associated with damage of the nerves responsible for functions

that regulate blood pressure, heart rate, bowel and bladder emptying, digestion, etc.

EDX findings:

•

NCS: Abnormal SNAPs and CMAPs

•

EMG: Abnormal activity

•

Most commonly presents with demyelination and axonal loss

Treatment: Rehabilitation, medications

AIDP, acute inflammatory demyelinating polyneuropathy; CIDP, chronic inflammatory demyelinating polyneuropathy;

CMAP, compound motor action potential; CSF, cerebrospinal fluid; EDX, electrodiagnostic; EMG, electromyography;

MSR, muscle stretch response; NCS, nerve conduction study; SNAP, sensory nerve action potential.430 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

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NEUROMUSCULAR JUNCTION DISORDERS

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These disorders hinder the production, release, or uptake of ACh at the NMJ. A low safety fac-

tor causes the amplitude of the endplate potentials to fall below the threshold needed to

generate a muscle fiber AP. This occurs due to an alteration of quantal response or content

(Table 5–43).

Myasthenia gravis (MG) is a disorder resulting in a decreased quantal response due to an

­ autoimmune response against postsynaptic ACh receptors. This leads to reduced miniature

endplate potential (MEPP) amplitudes, but their frequency remains normal (quantal content is

normal) (Figure 5–119).

Lambert–Eaton myasthenic syndrome (LEMS, myasthenic syndrome) is a disorder resulting in

decreased quantal content leaving the presynaptic cleft, resulting in normal MEPP amplitudes

but with decreased frequency (quantal response is normal).

ELECTRODIAGNOSTIC FINDINGS

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Evaluation of the NMJ involves typical NCS with EMG, as well as the addition of repetitive

nerve stimulations (RNSs) and single-fiber EMG (SFEMG; if needed;).

NCS:

– – SNAP: Typically normal. Occasionally, sensory neuropathies can be associated with a

­ paraneoplastic syndrome seen with LEMS.

– – CMAP: Normal or decreased amplitude. If low, this should be followed by a 10 second maxi-

mum voluntary contraction, followed by a single stimulation. Increased amplitudes >100%

compared with premaximum contraction are seen in LEMS.

EMG: Can be normal or abnormal

– – In severe cases, abnormal spontaneous activity can be seen. Short-duration, low-amplitude

MUAPs with early recruitment are associated with blocking.

•

– – – – MUAPs become unstable (variable amplitudes and configurations—Figure 5–120)

RNS and SFEMG:

Abnormal RNS and SFEMG studies. See next sections.

Control

NT

1 µ

Membrane

length

5.83 µ/µ2

Myasthenia gravis Myasthenic syndrome

3.95 µ/µ2

6.47 µ/µ2

FIGURE 5–119 Postsynaptic membrane changes. MG—

Simplification of the postsynaptic membrane. The NMJ

demonstrates a reduction in the number of postsynaptic

junctional folds. MS—Hypertrophy of postsynaptic

membrane; the NMJs demonstrate an increase in the

complexity of the postsynaptic membrane architecture.

MG, myasthenia gravis; MS, myasthenic syndrome; NMJ,

neuromuscular junction.

FIGURE 5–120 Unstable MUAP. Same MUAP with

varying amplitudes. This is seen in patients with MG

(myasthenia gravis); amplitude variations are from

neuromuscular junction blocking.

MG, myasthenia gravis; MUAP, motor unit action

potential.TABLE 5–43 Neuromuscular Junction Disorders

DISEASE MYASTHENIA GRAVIS LAMBERT–EATON SYNDROME BOTULISM

Location Postsynaptic Presynaptic (LEMS) Presynaptic

Etiology •

•

A disorder of neuromuscular transmission

due to an ­ autoimmune response in which

polyclonal antibodies are directed against the

Muscle Specific Tyrosine Kinase (MuSK) of

the postsynaptic membrane.

Associated with thymic disorder or thymic

tumor.

•

•

•

A disorder of neuromuscular transmission

due to an autoimmune response against the

active sites (voltage gated P/Q Ca channels

on the presynaptic membrane)

This decreases Ca++ entry into the cell,

causing a decreased release of ACh into the

synaptic cleft.

Associated with small cell (oat cell) carci-

noma of the lung (50% are paraneoplastic).

•

•

A disorder of ­ neuromuscular transmission

caused by Clostridium botulinum toxins

blocking presynaptic exocytosis of ACh from

the nerve terminal.

Associated with ingestion of contaminated

raw meat, fish, canned vegetables, and raw

honey.

Onset Bimodal distribution

First Peak: 20–30 years,

Female > male

Second Peak: 60–80 years,

Female = male

Bimodal distribution

First Peak: 40 years,

Female > male

Second Peak: 60 years,

Male > female

Begins 2–7 days after ingestion

Clinical presentation • Painless proximal fatigue and weakness.

Ocular weakness is the most common.

•

Exacerbated with ­ exercise, heat, or time of

day (evening)

•

•

Normal MSR

Facial or bulbar symptoms:

− Ocular weakness (Ptosis)

−

− Diplopia

−

− Dysphagia

−

− Dysarthria

−

•

•

Improved with rest

Edrophonium (Tensilon) Test: 2-mg dose fol-

lowed by an 8-mg dose, improvement begins

in 1 minute

•

•

•

•

•

•

•

•

Proximal fatigue and weakness

Mainly affects the lower limbs first

(quadriceps)

Abnormal MSR

Exacerbated with rest

Improved with exercise

Viselike grip

Rarely involves the neck, facial, or bulbar

muscles in contrast to MG

Autonomic symptoms:

− Dry mouth

−

− Erectile dysfunction

−

− Constipation

−

•

•

•

•

•

Decreased deep tendon reflexes

Bulbar symptoms are noted first:

− Ocular weakness (Ptosis)

−

− Dysphagia

−

− Dysarthria

−

GI symptoms:

− Diarrhea, N/V

−

− Widespread paralysis

−

or flaccidity

Abnormal MSR

Respiratory and cardiac dysfunction

NEUROMUSCULAR JUNCTION DISORDERS

(Continued )

431TABLE 5–43 Neuromuscular Junction Disorders (Continued)

DISEASE MYASTHENIA GRAVIS LAMBERT–EATON SYNDROME BOTULISM

Labs Muscle biopsy: Simplification of the postjunc-

tional membrane with loss of junctional folds

and receptors (Figure 5–119)

Antibody testing:

•

•

Anti-Ach receptor antibodies

Anti-MuSK antibodies

Muscle biopsy: Overdevelopment of

­ neuromuscular junction (Figure 5–119)

Decreased active zones are noted

Antibodies against voltage-gated Ca+ channels

Monitor for cancer

Botulinum toxin: Found in stool or blood serum

EDX findings NCS

•

Normal SNAP and CMAP

•

>10% decrement on low rate rep. stim.

EMG

•

•

Unstable MUAP, drop-off occurs with sus-

tained ­ contraction (Figure 5–120)

See single-fiber EMG

NCS

•

SNAP: Normal

•

CMAP: Low amplitude

•

>10% decrement on low rate rep. stim.

EMG

•

•

Unstable MUAP, drop-off occurs with sus-

tained contraction

See single-fiber EMG

NCS

•

SNAP: Normal

•

CMAP: Abnormal amplitude

•

>10% decrement on rep. stim. study

EMG

•

•

Unstable MUAP

See single-fiber EMG

Treatment •

•

•

•

•

•

•

Thymectomy

Anticholinesterase drugs:

− Mestinon (Pyridostigmine) 30 mg q 4 to

−

6 hours

Corticosteroids

Immunosuppressive agents

Plasmapheresis

One-third improve spontaneously

IV Immunoglobulin

•

•

•

•

•

•

•

Treat malignancy

Corticosteroids

Immunosuppressive agents

Plasmapheresis

Guanidine: Increases ACh quanta

− Side effects: GI, bone marrow suppres-

−

sion, renal tubular necrosis

3,4-diaminopyridine

IV Immunoglobulin

•

•

•

Treat with trivalent ABE antitoxin in first 24

hours

failure

Supportive with intubation for respiratory

Recovery occurs from ­ collateral sprouting

ACh, acetylcholine; CMAP, compound muscle action potential; EMG, electromyography; GI, gastrointestinal; LEMS, Lambert–Eaton myasthenic syndrome; MG, myasthenia gra-

vis; MSR, muscle stretch response; MUAP, motor unit action potential; NCS, nerve conduction study; N/V, nausea and vomiting; rep. stim., repetitive stimulation; SNAP, sensory

nerve action potential.

432 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGYNEUROMUSCULAR JUNCTION DISORDERS

433

REPETITIVE NERVE STIMULATION (RNS) (FIGURE 5–121 AND

TABLE 5–44)

•

These are studies in which a repeated

­ supramaximal stimulation of a motor nerve

is performed.

•

A series of CMAPs are recorded for

­ pathologic amplitude changes. Muscles

should be evaluated in a proximal

­ progression if an abnormality is suspected

but not demonstrated.

•

The study is best performed on the clini-

cally weak muscle(s). However, due to

the ease of the examination, it is typically

started in the hand intrinsics. If no abnor-

mality is noted, then progression to more

FIGURE 5–121 Repetitive nerve stimulation: normal response.

proximal muscles is performed.

•

Proper setup is essential to obtain the appropriate responses. Prior to starting the study, cholin-

esterase inhibitors should be held for 12 hours, if medically cleared.

TABLE 5–44 Muscle Evaluation for RNS

PROGRESSION MUSCLES

First ADM or APB

Second Deltoid

Third Trapezius

Fourth Orbicularis oculi

ADM, abductor digiti minimi; APB, abductor pollicis brevis; RNS, repetitive nerve stimulation.

RNS SETUP

•

Immobilize the electrode

•

Immobilize the limb

•

Stimulate at a supramaximal level

•

Optimize limb temperature (approximately 30°C)

•

Minimize electrode gel

•

Stop anticholinesterase inhibitors

ABNORMALITY

A >10% decrease in amplitude from the first to fifth waveform is significant for pathology

Low-Rate Repetitive Stimulation (Figure 5–122)

•

This repetitive stimulation test is performed at a rate of

2 to 3 Hz.

•

Each stimulus causes the EPP amplitude to drop. If the

safety factor is decreased, the potential will fall below

the threshold necessary for activation. This results in a

decrease of the MUAP amplitude (Table 5–45).

•

An abnormality is considered when a CMAP demon-

strates >10% amplitude reduction between the first and

fourth waveforms.

•

An increase in waveform can be seen if more stimula-

tions are provided due to mobilization of ­ secondary ACh

stores.

•

A typical U-shaped decrement can be seen in myasthenia gravis.

FIGURE 5–122 Low-rate repetitive

stimulation decremental response.434 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

TABLE 5–45 LRRS Amplitude Changes

DISORDER AMPLITUDE CHANGE

Myasthenia gravis >10% decrement

Lambert–Eaton syndrome >10% decrement

Botulism >10% decrement

LRRS, low-rate repetitive stimulation.

Postactivation Facilitation (PAF)

After a decrement is noted with LRRS, a 30- to 60-second isometric contraction or ­ tetany-producing

stimulation (50 Hz) should be performed. Postactive Facilitation (PAF) demonstrates a repair in

the CMAP amplitude with an immediate follow up LRRS because of an improvement in neuro-

muscular transmission.

CMAP, compound muscle action potential; LRRS, low-rate repetitive stimulation.

Postactivation Exhaustion (PAE)

This response is seen as a CMAP amplitude decreases. It occurs with a LRRS ­ performed every

minute for 5 minutes after an initial 30- to 60-second isometric contraction. The greatest drop off

is between 2 and 4 minutes. This test should be used if a decrement does not present with the

initial LRRS, but a diagnosis of a NMJ disorder is suspected (Figure 5–123).

CMAP, compound muscle action potential; LRRS, low-rate repetitive stimulation; NMJ, neuromuscular junction.

Rested

Muscle

REPETITIVE NERVE STIMULATION

NORMAL (N), MYASTHENIA GRAVIS (MG),

LAMBERT – EATON MYASTHENIC SYNDROME (LEMS)

Exercise

Time

Time After Exercise

3 s

2 min

10 min

N

30 s

–

5 mV

+

10 ms

MG

30 s

LEMS

10 s

–

5 mV

+

10 ms

–

5 mV

+

7 ms

FIGURE 5–123 Repetitive stimulation (a decrement must be reproducible on a number of trials).NEUROMUSCULAR JUNCTION DISORDERS

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High-Rate Repetitive Stimulation (Figure 5–124, TABLE 5–46)

•

This repetitive stimulation test is performed at a rate of 10 to 50 Hz. It causes an accumulation

of calcium in the cell, which assists ACh release and repairs the waveforms.

•

High-rate repetitive stimulation (HRRS) is uncomfortable and is typically performed if a patient

is unable to perform a 30- to 60-second maximal isometric contraction.

FIGURE 5–124 High-rate repetitive stimulation. (I) Increment with 50 Hz stimulation. (II) Increment with voluntary

contraction (50 Hz simulation/train of 50, femoral/rectus femoris, 500% facilitation).

TABLE 5–46 High-Rate Repetitive Stimulation Amplitude Changes

DISORDER AMPLITUDE CHANGE

Myasthenia gravis Decrement demonstrated and partially

repaired

Lambert–Eaton syndrome 200%–300% increment

Botulism Mild increment

Pseudofacilitation (Figure 5–125)

•

This is a normal reaction and demonstrates a progressive increase in CMAP amplitude with

HRRS or voluntary muscle contraction.

•

It represents a decrease in temporal dispersion due to increased synchronicity of muscle fiber

­ contraction. The waveforms produced maintain a constant area under the curve though the

­ amplitude appears increased because the duration is decreased.436 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

–

4 mV

+

2 ms

FIGURE 5–125 Pseudofacilitation. Repetitive nerve stimulation study in a normal subject. The successive M waves

were recorded with surface electrodes over the hypothenar eminence (abductor digiti quinti) during ulnar nerve

stimulation at a rate of 30 Hz. Pseudofacilitation may occur in normal subjects with repetitive nerve stimulation

at high (20–50 Hz) rates or after strong volitional contraction, and probably reflects a reduction in the temporal

dispersion of the summation of a constant number of muscle fiber action potentials due to increases in the

propagation velocity of action potentials of muscle cells with repeated activation. Pseudofacilitation should be

distinguished from facilitation. The recording shows an incrementing response characterized by an increase in the

amplitude of the successive M waves with a corresponding decrease in the duration of the M wave resulting in no

change in the area of the negative phase of the successive M waves.

RNS FINDINGS IN NMJ DISORDERS

NMJ DISORDER

TEST MYASTHENIA GRAVIS LEMS BOTULISM

CMAP amplitude Normal or reduced Decreased Decreased

RNS >10% decrement noted

between first and fourth to fifth

stimulation

>10% decrement in

amplitude

>10% decrement in

­ amplitude, or variable

changes

PAF 20%–50% improvement >100% improvement >40% improvement

PAE Observed 2–4 minutes after

maximal voluntary contraction

A train of five stimuli

every minute for 5

minutes to monitor for

decrease

Absent

CMAP, compound muscle action potential; LEMS, Lambert–Eaton myasthenic syndrome; NMJ, neuromuscular

junction; PAE, postactivation exhaustion; PAF, post activation facilitation; RNS, repetitive nerve stimulation.

SINGLE-FIBER EMG

•

This is a study that monitors the parameters of single muscle fiber APs. It is useful if repetitive

stimulation of at least three muscles is normal and an abnormal diagnosis is still suspected.

•

•

SFEMG is the most sensitive test for NMJ disorders but has low specificity.

Abnormalities can be associated with NMJ disorders, motor neuron disorders, and peripheral

neuropathies.

PARAMETERS

•

Fiber density (FD; Figure 5–126):

– – This represents the number of single fibers belonging to the same motor unit within the

recording radius of the electrode. The FD is determined by dividing the number of single

muscle fiber APs at 20 sites by 20.

– – A FD of 1.5 is normal. Higher than this represents a denervation and reinnervation process.MYOPATHIES

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•

•

Jitter (Figure 5–127):

– – During voluntary contraction a small variation exists

between the interpotential discharges of two muscle

fibers belonging to the same motor unit. This varia-

tion is normally 10 to 60 μsec. It is ­ typically consid-

ered abnormal if it is longer than this.

– Disorders of neuromuscular transmission affect the

–

safety factor and cause a delay in the time for an EPP to

reach threshold for a muscle fiber AP, which increases

the jitter between the two neighboring muscle fibers.

Reinnervation through collateral sprouting after a nerve

injury also can cause a delay. The immature NMJs have

poor activation, resulting in increased jitter within the

first month.

– – This is seen in conditions including amyotrophic lat-

eral sclerosis (ALS), NMJ disorders, axonal neuropa-

thies, and myopathies.

Blocking:

– This is an abnormality that occurs when a single muscle

–

fiber AP fails to appear. It occurs if the jitter becomes

>100 μsec. It typically resolves in approximately 1 to 3

months, after reinnervation is completed. However, the

increased jitter may take approximately 6 months to

resolve.

Normal

**A**

Reinnervation

**B**

FIGURE 5–126 Increased fiber density. The

dots represent single muscle fibers of one

motor unit with the recording radius.

(A) Normal muscle (action potentials from

1 to 2 fibers recorded). (B) Reinnervation

(action potentials from many fibers

recorded).

FIGURE 5–127 Single-fiber EMG recordings. Top: Superimposed view. Bottom: Rastered view. (A) Normal. (B)

Increased jitter. (C) Increased jitter with blocking.

EMG, electromyography.

n

MYOPATHIES

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These are skeletal muscle fiber disorders that can occur from a variety of etiologies.

Important factors to consider in its diagnosis include age of onset, developmental milestones,

­ familial involvement, prodromal illness, and patient history.

Currently genetic testing has demonstrated a greater ability to classify the type of myopathy.

Please refer to the pediatric section for additional information on this topic.438 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

ETIOLOGY (TABLE 5–47)

TABLE 5–47 Etiology of Myopathies

DYSTROPHIC CONGENITAL METABOLIC INFLAMMATORY ENDOCRINE TOXIC STEROID

•

•

•

•

•

Duchenne

Becker

Limb-girdle

disorder

Facioscapulohumeral

Myotonic

•

•

•

•

Central core

Nemaline rod

Centronuclear

Fiber type

disproportion

•

•

•

•

•

Acid maltase

deficiency

Myophosphorlyase

deficiency

Phosphofructokinase

deficiency

Hyperkalemic

­ periodic

paralysis

Hypokalemic

­ periodic

paralysis

•

•

•

•

•

•

Polymyositis/

dermatomyositis

Sarcoidosis

Viral

Bacterial

Parasitic

Inclusion body

myositis

•

•

Thyroid

parathyroid

Adrenal

pituitary

•

•

•

•

Alcohol

Diuretic

Vincristine

Steroid

•

Corticosteroid

use

The Role of Dystrophin

•

Dystrophin is a protein found in the sarcolemma of normal muscle. It provides mechanical

support and structural integrity for the muscle membrane cytoskeleton.

•

Mutation in the dystrophin gene leads to muscle fiber necrosis. Patients present with clinical

symptoms of myalgias, fatigue, and weakness.

•

Muscle biopsies help differentiate between dystrophinopathies. In Duchenne ­ muscular

dystrophy, dystrophin is absent or markedly deficient. In Becker’s ­ muscular dystrophy, the

abnormalities are less severe.

CLINICAL PRESENTATION

•

•

•

The patient may demonstrate muscle-related changes presenting as atrophy, hypertrophy,

abnormal MSR, weakness, hypotonia, gait abnormalities, or myotonia.

Myotonia is a painless delayed relaxation of skeletal muscles following a voluntary contraction.

It is exacerbated by cold but relieved with exercise, Dilantin, procainamide, and calcium

channel blockers.

Arthrogryposis, which is a fixed deformity of the extremities due to intrauterine hypomobility,

may occur in newborns from myopathies, muscular dystrophies, or oligohydramnios.

A hallmark sign of myopathy is the inability to generate a forceful contraction.

ELECTRODIAGNOSTIC FINDINGS

NCS

•

SNAP: Normal

•

CMAP: Decreased amplitude with significant muscle fiber atrophy. Normal latencies and con-

duction velocities.

EMG

•

Classic findings are low amplitude, short duration, polyphasic MUAP with early recruitment

(Tables 5–48).

•

Resting activity: Abnormal activity depends on the type of disorder involved (Tables 5–49).

Quantitative EMG

•

This study may provide a more detailed measurement of the MUAPs. It is a better indication

of waveform duration, which is a sensitive parameter for diagnosing myopathies. The mean

­ duration is calculated using 20 MUAPs and on a screen set with a trigger and delay line. This

avoids ­ superimposing MUAPs and falsely creating a polyphasic.MYOPATHIES

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TABLE 5–48 Recruitment: Early Onset With Minimal Effort

PRESENTATION POSSIBLE CAUSES OF MUAP ALTERATIONS

SDSA These classic polyphasic potentials are due to loss of muscle fibers.

LDLA These polyphasic potentials are due to collateral sprouting.

Unstable These variable amplitude potentials are due to blocking of immature NMJs, which are

formed at the beginning of collateral sprouting.

LDLA, long-duration, large amplitude; NMJ, neuromuscular junction; SDSA, short-duration, small amplitude.

TABLE 5–49 Abnormal Spontaneous Activity in Myopathies

FIBRILLATIONS AND

POSITIVE SHARP WAVES

COMPLEX REPETITIVE

DISCHARGE MYOTONIC DISCHARGE

•

•

•

•

•

•

•

•

•

•

•

•

•

Polymyositis

Dermatomyositis

Inclusion body myopathy

Trichinosis

Toxic myopathies

Direct muscle trauma

Rhabdomyolysis

Acid maltase deficiency

Myotubular myopathy

Hyperkalemic periodic paralysis

Nemaline rod

Sarcoid myopathy

Muscular dystrophies

•

•

•

•

•

Polymyositis

Dermatomyositis

Muscular dystrophies

Schwartz-Jampel syndrome

Inclusion body myopathy

•

•

•

•

•

•

•

•

•

•

•

Myotonia congenita

Myotonic dystrophy

Paramyotonia congenita

Hyperkalemic periodic paralysis

Acid maltase deficiency

Hypothyroid myopathy

Myotubular myopathy

Chloroquine myopathy

Diazocholesterol intoxication

Polymyositis

Dermatomyositis

Repetitive Nerve Stimulation

•

A normal or a decremental response can occur. This is due to the reduced safety factor found in

regenerating immature NMJs that form during recovery or reinnervation.

Single-Fiber EMG

•

This can demonstrate increased jitter, FD, and blocking.

Additional Testing: Muscle Biopsy

TYPE I FIBER ATROPHY TYPE II FIBER ATROPHY

•

•

•

Myotonic dystrophy

Nemaline rod myopathy

Fiber type disproportion

•

•

•

Steroid myopathy

Myasthenia gravis

Deconditioning

TYPES OF MYOPATHIES

•

•

The following tables outline pertinent myopathic patterns.

Please refer to Table 5–47 as an overview for Tables 5–50 through 5–56.TABLE 5–50 Dystrophic Myopathies: Common Disorders

DISORDER

Etiology X-linked recessive (xp21), spontaneous X-linked recessive Autosomal dominant Autosomal dominant

Onset 3–5 years old Adulthood Infant Childhood-early adult

Course Severely progressive (death by 20s) Slowly progressive Clinical

presenta-

tion

DUCHENNE MUSCULAR DYSTROPHY (MOST

COMMON MYOPATHY)

•

•

•

•

Proximal weakness

Calf pseudohypertrophy

Cardiomyopathy

Less mental retardation

than DMD

•

•

•

•

•

•

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•

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BECKER MUSCULAR

DYSTROPHY

Proximal muscle weakness (pelvic girdle)

Abnormal MSR

Increased lumbar lordosis

Ambulation difficulties: Toe walking (<5 years), clumsy

running (<7 years)

Gower’s sign: Difficulty rising from the floor due to hip

and knee extensor weakness

Calf pseudohypertrophy with fat and fibrous tissue

Contractures: Iliotibial band (first), Achilles tendon

Scoliosis, causing cardiomyopathy and restrictive lung

disease

Possible mental retardation

Wheelchair by 12 years old

Extraocular muscles are spared

MYOTONIC DYSTROPHY (STEINERT’S;

SECOND MOST COMMON)

Spreads to other muscles

FACIOSCAPULOHUMERAL

(FSH) DYSTROPHY

•

•

•

•

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•

Weakness: Distal > proximal myotonia with

sustained grip

Hatchet face (wasting of the temporalis and

masseter)

Frontal balding

Poor vision

Ptosis

Impotence

Hypertrichosis

Mental retardation

Cardiac abnormalities

Endocrine abnormalities

Congenital myotonic dystrophy:

− “Shark mouth” appearance

−

Facial diplegia

Possible club foot

•

•

•

•

•

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•

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•

Proximal muscle weakness

Facial droop

Weak eye closing

Weak forehead wrinkling

Arm atrophy with deltoid and forearm

sparing (Popeye arm)

Cataracts (dry sclera)

Retinopathy

Lip protrusion

Transverse smile

Frontal balding

Testicular atrophy

Extraocular muscles are spared

#1 muscle to test in FSH is tibialis

anterior

Inability to whistle

Labs M Bx: No dystrophin, internal nuclei variation in fiber size.

Blood: Increased CPK and aldolase. ECG: Abnormal

M Bx: Decreased dystrophin

(15%–85%), increased CPK

M Bx: Type I fiber atrophy with Type II hypertro-

phy. No dystrophin involvement

M Bx: Scattered fiber necrosis and

regeneration. Inflammatory infiltrate may

be noted

EDX

findings

NCS

•

•

EMG

•

SNAP: Normal

CMAP: ± Decreased amplitude

AA (rare), ER, ± SDSA MUAP

NCS

•

•

EMG

•

SNAP: Normal

CMAP: ± Decreased amplitude

AA (rare), ER, SDSA MUAP

NCS

•

•

EMG

•

SNAP: Normal

CMAP: ± Decreased amplitude

AA (rare), ER, SDSA MUAP, myotonia

NCS

•

•

EMG

•

SNAP: Normal

CMAP: Decreased amplitude in the

involved muscles

AA, ER, SDSA MUAP

Treatment Rehabilitation. Scoliosis surgery before the vital capacity

is below 35% (usually due to a curve of >30 degree).

Prednisone

Rehabilitation: Bracing, tendon

lengthening, possible scoliosis

surgery

Rehabilitation: bracing, medications: procain-

amide, Dilantin, and quinine (PDQ). May need a

pacemaker

Rehabilitation

AA, abnormal activity; CMAP, compound muscle action potential; CPK, creatine phosphokinase; DMD, Duchenne muscular dystrophy; EMG, electromyography; ER, early recruit-

ment; FSH, facioscapulohumeral; M Bx, muscle biopsy; MSR, muscle stretch response; MUAP, motor unit action potential; NCS, nerve conduction study; SDSA, short duration,

small amplitude; SNAP, sensory nerve action potential.

440 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGYTABLE 5–51 Congenital Myopathies: Common Disorders

DISORDER CENTRAL CORE DISEASE NEMALINE ROD MYOPATHY

CENTRONUCLEAR

MYOTUBULAR

FIBER TYPE

DISPROPORTION

Genetic Etiology Autosomal dominant Autosomal dominant/recessive X-linked recessive Variable

Onset Infancy Infancy Infancy Infancy

Clinical presentation •

•

•

•

•

Floppy infant/hypotonia

Proximal weakness

Congenital hip dislocation

Delayed milestones

Associated with malignant

hyperthermia

•

•

•

•

•

•

•

•

Floppy infant/hypotonia

Diffuse weakness

Facial involvement

Narrowed long face

High arched palate

Death: Respiratory failure

Foot drop

EOM spared

•

•

•

•

•

•

Floppy infant/hypotonia

Ptosis

Extra ocular muscle

involvement

Facial diplegia

Dysphagia

Respiratory insufficiency

•

•

•

Floppy infant/hypotonia

Hip contractures

Hip dislocations

Labs M Bx: Central cores in Type I

fibers. Absent mitochondria

M Bx: Rod-shaped bodies on

Gomori trichrome stain

M Bx: Central ­ location of fiber

nuclei, forming chains

M Bx: Numerous small Type I

and ­ normal to large Type II fibers

EDX findings EMG

•

ER, SDSA MUAP

EMG

•

ER, SDSA MUAP

EMG

•

AA, ER, SDSA MUAP

EMG

•

ER, SDSA MUAP

Treatment Bracing Rehabilitation, surgery Rehabilitation, anti-seizure

Rehabilitation, bracing

medication

AA, abnormal activity; EDX, electrodiagnostic; EMG, electromyography; EOM, extraocular muscles; M Bx, muscle biopsy; MUAP, motor unit action potential; SDSA, short

­ duration, small amplitude.

MYOPATHIES

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TABLE 5–52 Inflammatory Myopathies

DISORDER POLYMYOSITIS/DERMATOMYOSITIS INCLUSION BODY MYOSITIS

Etiology Autoimmune, connective tissue disorder, infec-

tion, cancer

Unknown

Clinical presentation • Symmetrical proximal weakness: Hips fol-

lowed by shoulders

•

Neck flexion weakness

•

Myalgias, dysphagia, dysphonia

•

No facial or ocular muscle weakness

•

Dermatomyositis:

Periorbital violet rash and edema

Gottron’s sign: Red-purple patches over the

knuckles, elbows, knees

•

•

•

Asymmetric, slowly progressive,

painless weakness in proximal

and distal muscles

Associated with a

polyneuropathy

Affects adults 45–55 years and

peaks at 70 years

Labs Blood: Increased CPK, ESR, aldolase, SGOT,

SGPT, LDH

M Bx: Necrosis of the Type I and II fibers.

Perifascicular atrophy

Blood: Increase in CK

M Bx: Rimmed or cytoplasmic/basophilic

vacuoles. Eosinophilic

inclusion bodies

EDX findings NCS

•

•

EMG

•

SNAP: Normal

CMAP: Normal

AA (most commonly in the paraspinals)

ER, SDSA MUAP

NCS

•

•

EMG

•

SNAP: ± Abnormal

CMAP: ± Abnormal

AA, ER, ± SDSA MUAP

Treatment Rehabilitation: Corticosteroids, ­ cytotoxic agents,

IV Ig, ­ plasmapheresis, rest.

Hydroxycholoroquine for skin ­ manifestations

(dermatomyositis)

Rehabilitation: This condition is

refractory to steroid treatment. No

treatment.

AA, abnormal activity; CMAP, compound muscle action potential; CPK, creatine phosphokinase; EDX, electro-

diagnostic; EMG, electromyography; ER, early recruitment; ESR, erythrocyte sedimentation rate; LDH, lactate

dehydrogenase; M Bx, muscle biopsy; MUAP, motor unit action potential; NCS, nerve conduction study; SDSA,

short duration, small amplitude; SGOT, serum glutamic oxaloacetic transaminase; SGPT, serum glutamic pyruvic

transaminase; SNAP, sensory nerve action potential.

TABLE 5–53 Metabolic Myopathies: Common Disorders

CHARACTERISTICS MCARDLE’S DISEASE (TYPE V) POMPE’S DISEASE (TYPE II)

Etiology Autosomal recessive

Myophosphorylase deficiency

Autosomal recessive

Acid maltase deficiency

Onset <15 years of age Infant to adult

Clinical presentation • Exercise intolerance

•

•

•

•

Easy fatigability

Muscle stiffness

Cramping

Second-wind phenomenon: Brief rest

improves symptoms

•

Strenuous exercise can precipitate myolysis

(possibly cause renal failure and death)

•

•

•

•

•

•

•

Hypotonia

Tongue enlargement

Cardiomegaly

Hepatomegaly

Respiratory insufficiency

Death by 2 years of age

A milder form may affect adults

(Continued )MYOPATHIES

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TABLE 5–53 Metabolic Myopathies: Common Disorders (Continued)

CHARACTERISTICS MCARDLE’S DISEASE (TYPE V) POMPE’S DISEASE (TYPE II)

Labs Urine: Myoglobinuria

M Bx: Excess glycogen, absent phosphorylase

Blood: Increase CK during the

attacks M Bx: Vacuoles in Type I and

II fibers

EDX findings NCS

•

•

EMG

•

SNAP: Normal

CMAP: Normal

Electrical silence during attacks

(contracture)

NCS

•

•

EMG

•

SNAP: Normal

CMAP: Normal

AA, ER, SDSA MUAP

Treatment Supportive Supportive

AA, abnormal activity; CK, creatine kinase; CMAP, compound muscle action potential; EDX, electrodiagnostic; EMG,

electromyography; ER, early recruitment; M Bx, muscle biopsy; MUAP, motor unit action potential; NCS, nerve

conduction study; SDSA, short duration, small amplitude; SNAP, sensory nerve action potential.

TABLE 5–54 Metabolic Myopathies Periodic Paralysis: Common Disorders

CHARACTERISTICS

HYPERKALEMIC PERIODIC

PARALYSIS HYPOKALEMIC PERIODIC PARALYSIS

Etiology Autosomal dominant

Multiple secondary causes

Autosomal dominant

Multiple secondary causes

Onset Childhood–second decade Starts in early second decade

Clinical presentation • Proximal muscle weakness

•

Paresthesias of the lips and lower

limbs

•

Myotonia

•

Attacks last 10–60 minutes

•

May be aborted with exercise

•

Exacerbated with cold exposure and

rest following exercise

•

•

•

•

Weakness starts in the legs and spreads

proximally

Attacks last 12–24 hours

Myotonia seen in the eyelids

Exacerbated with rest after exercise,

stress, and a high carbohydrate diet

Labs Blood: High K+ during the attack Blood: Low potassium

M Bx: Normal

EDX findings NCS

•

•

SNAP: Normal

CMAP: Normal

EMG

•

During an attack: ER, SDSA

MUAP , AA

NCS

•

•

EMG

•

SNAP: Normal

CMAP: Normal

During an attack: Electrical silence

Treatment Diet: High carbohydrate Diet: K+ supplement

AA, abnormal activity; CMAP, compound muscle action potential; EDX, electrodiagnostic; EMG, electromyogra-

phy; ER, early recruitment; M Bx, muscle biopsy; MUAP, motor unit action potential; NCS, nerve conduction study;

SDSA, short dURATIOn, small amplitude; SNAP, sensory nerve action potential.444 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

Table 5–55 Myopathies: Common Disorders

CHARACTERISTICS

MYOTONIA CONGENITA

(THOMSEN’S DISEASE; LITTLE

HERCULES)

PARAMYOTONIA CONGENITA

(EULENBURG)

Etiology Autosomal dominant Autosomal dominant

Onset Birth–adulthood Birth–adulthood

Clinical presentation • Severe spasms exacerbated by the cold

•

Improves with warmth and exercise

•

Muscle hypertrophy

•

Myotonia

•

No weakness

•

•

•

•

•

Stiffness

Weakness

Fatigue

Myotonia

Exacerbated with cold and exercise

Labs Blood: CK—normal M Bx: Fiber size variation

EDX findings NCS

•

•

EMG

•

SNAP: Normal

CMAP: Normal

AA (myotonic discharges, no fibs, no pos.

waves), normal recruitment, normal MUAP

NCS

•

SNAP: Normal

•

CMAP: Decreased with cooling

EMG

•

Electric silence or AA with cooling.

Treatment Medication: Procainamide, Dilantin, quinine (PDQ) Warm extremities

AA, abnormal activity; CK, creatine kinase; CMAP, compound motor action potential; EDX, electrodiagnostic; EMG,

electromyography; M Bx, muscle biopsy; MUAP, motor unit action potential; NCS, nerve conduction study; SNAP,

sensory nerve action potential.

TABLE 5–56 Steroid Myopathy

CHARACTERISTICS STEROID MYOPATHY STATIN MYOPATHY

Etiology Corticosteroid proteolysis effect Multifactorial

Onset Weeks to years post use Weeks to years post use

Clinical presentation • Proximal muscle weakness

•

•

Risk is increased if on 30 mg/day

Preferentially affects the hip girdle

muscles

•

•

•

Proximal weakness and pain

Worse with exercise

Lipophilic statins (simvastatin, ator-

vastatin, lovastatin) are more likely

to produce muscular effects than are

relatively hydrophilic agents (pravas-

tatin, rosuvastatin, fluvastatin)

Labs M Bx: Type II atrophy M Bx: Nonspecific

EDX findings NCS

•

•

EMG

•

SNAP: Normal

CMAP: Normal

May be abnormal with small

­ polyphasics in very severe cases

NCS

•

•

EMG

•

SNAP: Normal

CMAP: Normal

May be abnormal with small

­ polyphasics in very severe cases

Treatment Rehabilitation, exercise, stop steroids Stop statins, or stop medications that

interfere with Cy-P450: macrolide antibi-

otics, verapamil, diltiazem, cimetidine, etc.

CMAP, compound motor action potential; EDX, electrodiagnostic; EMG, electromyography; M Bx, muscle biopsy;

NCS, nerve conduction study; SNAP, sensory nerve action potential.MOTOR NEURON DISEASE

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MOTOR NEURON DISEASE

•

•

This is a disorder resulting from the progressive degeneration of the motor neurons in the spi-

nal cord, brainstem, or motor cortex (Tables 5–57).

It manifests as muscular weakness and atrophy with varying corticospinal tract signs. See pedi-

atric section for further information.

ETIOLOGY

TABLE 5–57 Motor Neuron Diseases

LOWER MOTOR NEURON

LESION

UPPER AND LOWER MOTOR

NEURON LESION

UPPER MOTOR NEURON

LESION

SMA

Poliomyelitis/Post-polio syndrome

ALS PLS

Hereditary spastic paraplegia

ALS, amyotrophic lateral sclerosis; PLS, primary lateral sclerosis; SMA, spinal muscle atrophy.

CLINICAL PRESENTATION

TABLE 5–58 Lower Versus Upper Motor Neuron Signs

LOWER MOTOR NEURON SIGNS UPPER MOTOR NEURON SIGNS

Atrophy

Flaccidity

Hyporeflexia

Fasciculations

Weakness

Spasticity

Hyperreflexia

Upgoing plantar response

EDX FINDINGS

•

Evaluation should include at least one upper and one lower limb, starting in the most severely

affected muscles. It is important to rule out treatable neuropathies that mimic motor neuron

disease (MND). An example of this is multifocal motor neuropathy, which includes conduction

block and temporal dispersion, not seen in MND.

NCS

– – SNAP: Typically normal. Abnormal can be seen in hereditary spastic paraplegia and spino-

bulbar muscular atrophy.

– – CMAP: Variable findings

nn Abnormalities in weakened muscles with asymmetric side-to-side comparisons demon-

strating decreased CV and prolonged latencies.

– – F-wave: Abnormal latency, persistence with chronodispersion

EMG

– – – – – – – – Classic findings are seen in the area of clinical UMN and lower motor neuron (LMN) abnor-

malities. Active denervation with reinnervation must be found in three of four body seg-

ments (craniobulbar, cervical, thoracic, and lumbosacral). At least two muscles with different

innervations should be abnormal.

Chronic neurogenic changes (decreased MUP recruitment, large amplitude MUAP, rapidly

firing potential) and denervation potentials (FIB, PSWs, fasciculations) should be evident.

Recruitment is decreased. There are polyphasic potentials, due to reinnervation.

SFEMG: Abnormal fiber density, jitter, and blocking.446 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

TABLE 5–59 Abnormal Spontaneous Activity in Motor Neuron Disease

FIBs AND PSWs FASCICULATIONS CRDs

•

•

•

•

•

SMA Type I

SMA Type II

SMA Type III

ALS

Poliomyelitis

•

•

•

ALS

Poliomyelitis

Post-polio syndrome

•

SMA Type III

ALS, amyotrophic lateral sclerosis; CRD, complex repetitive discharge; FIB,fibrillation potential; PSW, positive sharp

waves; SMA, spinal muscular atrophy.

Recruitment of the Diaphragm

A monopolar needle recording electrode is inserted through the eighth or ninth ­ intercostal space

at the anterior axillary line. MUAP of the intercostal muscles is recruited during expiration and

is higher in amplitude than the diaphragm, which is recruited during inspiration.

MUAP, motor unit action potential.

The following tables outline pertinent MND patterns. Please refer to Table 5–57 as an overview for

Tables 5–60 and 5–61.

TABLE 5–60 Motor Neuron Diseases: Spinal Muscular Atrophy Types I, II, and III

CHARACTERISTICS

SPINAL MUSCULAR

ATROPHY TYPE I

(WERDNIG-HOFFMAN

DISEASE)

SPINAL MUSCULAR

ATROPHY TYPE II

SPINAL MUSCULAR

ATROPHY TYPE III

(KUGELBERG–

WELANDER DISEASE)

Genetic Etiology Autosomal recessive Autosomal recessive Autosomal recessive/

dominant

Onset 3–6 months 2–12 months 2–15 years

Course Death by 2–3 years of age Death by approximately

10 years old

Normal life expectancy

Worst prognosis Wheelchair by 2–3 years

of age

Wheelchair by 30 years

of age

Progression Rapid; fatal (respiratory failure) Slower; fatal (respiratory

failure)

Slowly

Clinical presentation • Floppy baby/hypotonia

•

Unable to reach milestones

•

Progressive weakness

•

Absent MSR

•

Difficulty feeding

•

Weak cry

•

Frog-legged position

•

Tongue fasciculations

•

Facial muscle affected

least

•

Extraocular muscles intact

•

Sphincter muscles are spared

•

Paradoxical breathing

•

Never sits independently

•

•

•

•

•

•

•

•

•

•

Floppy baby/hypotonia

Gradual progressive

limb weakness; upper

> lower

Absent MSR

Face least affected

Kyphoscoliosis

Equinus deformity of

the feet

± Tongue fasciculations

Progressive pulmonary

involvement

Independent sitting

Assistive devices for

standing and walking

•

•

•

•

•

•

•

•

•

Symmetric weakness:

Lower limb then upper

limb

Abnormal MSR

± Gowers’ sign

± Calf

pseudohypertrophy

± Dysphagia

± Dysarthria

Tongue

fasciculations—late

onset

Normal intelligence

Independent

standing/walking

(Continued )TABLE 5–60 MOTOR NEURON DISEASE

Motor Neuron Diseases: Spinal Muscular Atrophy Types I, II, and III (Continued)

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CHARACTERISTICS

SPINAL MUSCULAR

ATROPHY TYPE I

(WERDNIG-HOFFMAN

DISEASE)

SPINAL MUSCULAR

ATROPHY TYPE II

SPINAL MUSCULAR

ATROPHY TYPE III

(KUGELBERG–

WELANDER DISEASE)

Labs Blood: Increase CPK levels

M Bx: Hyper/atrophic fibers

Blood: Increase CPK levels

M Bx: Hyper/atrophic

fibers

Blood: Increase CPK

levels

M Bx: Hyper/atrophic

fibers

EDX findings NCS

•

•

EMG

•

SNAP: Normal

CMAP: ± Abnormal

AA, LDLA/SDSA MUAP, DR

NCS

•

•

EMG

•

SNAP: Normal

CMAP: ± Abnormal

AA, SDSA MUAP, DR

NCS

•

•

EMG

•

DR

SNAP: Normal

CMAP: Normal

AA, LDLA/SDSA MUAP,

Treatment Supportive Supportive

Rehabilitation

Supportive

Rehabilitation

AA, abnormal activity; CMAP, compound muscle action potential; CPK, creatine phosphokinase; DR, delayed

­ recruitment; DTRs, deep tendon reflexes; EDX, electrodiagnostic; EMG, electromyography; LDLA, long duration,

large amplitude; M Bx, muscle biopsy; MSR, muscle stretch response; MUAP, motor unit action ­ potential; NCS,

nerve conduction study; SDSA, short duration, small amplitude; SNAP, sensory nerve action potential.

TABLE 5–61 Motor Neuron Disease: ALS, Polio, Post-Poliomyelitis

CHARACTERISTICS ALS POLIOMYELITIS

POST-POLIOMYELITIS

SYNDROME

Pathology Degeneration of the anterior horn

cell

Degeneration of the

anterior horn cell

Loss of the anterior

horn cell

Etiology Unknown Picornavirus orally enters

the body and spreads via

lymphoid system leading to

orphaned muscle fibers

Death of the motor neu-

ron due to aging Burnout

of motor unit from

increased metabolic

demand (Figure 5–128)

Clinical presentation •

•

•

•

•

•

•

•

Most commonly in men after

the sixth decade

First signs: Asymmetric atro-

phy, weakness, fasciculations

Dysphagia (oral, pharyn-

geal), dysarthria

Pseudobulbar signs: A cluster

of symptoms including difficulty

chewing, swallowing, and

speech along with unprovoked

emotional outbursts (e.g., crying

and laughing)

Bowel and bladder are

spared

Sensation is spared

Extraocular muscles are spared

Upper and lower motor neuron

signs

•

•

•

•

•

•

•

Signs of infection: Fever,

malaise, sore throat,

vomiting, headache,

back and neck pain, and

stiffness

Weakness

Absent MSR

Bulbar palsies:

Dysphasia, nasal voice

Sensation is spared

Autonomic dysfunction

can occur

Prognosis: Disease can

progress or remit

− 25%: Severe disability

−

− 25%: Mild disability

−

− 50%: Complete

−

recovery

Halstead-Ross Criteria

1.

History of a previous

diagnosis

2.

Recovery of function

3. Stability for approxi-

mately 15 years

4.

Return of symptoms

5. No other medical

problems to explain

new symptoms:

− Weakness

−

− Atrophy

−

6. Difficulties with ADLs

− Fatigue

−

− Arthralgia

−

− Myalgia

−

− Cold intolerance

−

(Continued )448 5. ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

TABLE 5–61 Motor Neuron Disease: ALS, Polio, Post-Poliomyelitis (Continued)

CHARACTERISTICS ALS POLIOMYELITIS

POST-POLIOMYELITIS

SYNDROME

Clinical presentation

(cont.)

•

•

•

Prognosis: 50% die within

3 years, 30% live for

5 years, 10% live for

10 years

Wheelchair by 12–18 months

Predictors of survival:

− Age of onset (younger is

−

better)

− Severity of onset

−

− Pulmonary function

−

− Abnormal sniff test

−

correlates with poor

survival

•

Mortality: 1%–4%

chance in children. 10%

chance in adults with

bulbar and respiratory

involvement

EDX findings NCS

•

•

SNAP: Normal

CMAP: Normal

EMG

•

AA, DR, LDLA MUAP , CRDs

LRRS

•

Increased decrement

SFEMG

•

Increased jitter and fiber

density

Protocol:

Abnormal activity in two muscles

from two ­ different nerve roots in

three different body regions

Body regions:

Brainstem, cervical, thoracic,

lumbar

NCS

•

SNAP: Normal

•

CMAP: Normal or

decreased

EMG

•

AA, DR, LDLA MUAP

NCS

•

SNAP: Normal

•

CMAP: Abnormal

EMG

•

AA, DR, GIANT

MUAP

LRRS

•

Normal activity

SFEMG

•

Increased jitter,

fiber density, and

blocking

Post poliomy-

elitis syndrome

Electrophysiologically

resembles old

stable poliomyelitis.

Its diagnosis is not

based on EMG/NCS

but on clinical

presentation.

Treatment Rehabilitation, prevent

contractures, ­ submaximal

exercise, tracheostomy,

respiratory therapy,

riluzole (Rilutek®)

antiglutamate slows

disease ­ progression,

prolongs ventilator time,

BiPAP

Rehabilitation, pain

management, prevent

contractures

Rehabilitation, assis-

tive devices, energy

­ conservation, psycho-

logical counseling,

avoid fatigue

AA, abnormal activity; ADLs, activities of daily living; ALS, amyotrophic lateral sclerosis; CMAP, compound muscle

action potential; CRD, complex repetitive discharge; DR, delayed recruitment; EDX, electrodiagnostic; EMG, elec-

tromyography; LDLA, long duration, large amplitude; LRRS, low-rate repetitive stimulation; MSR, muscle stretch

response; MUAP, motor unit action potential; NCS, nerve conduction studies; SNAP, sensory nerve action potential;

SFEMG, single-fiber electromyography.WEAKNESS: DIFFERENTIAL DIAGNOSIS

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**Normal**

A B C D

**Acute polio**

Neuron that will survive

Dying neuron

A B C D

**Recovery and continuous remodeling PPMA**

Stable post-polio Early disease Late disease

A A B D

B

D

A B D

FIGURE 5–128 Post poliomyelitis progressive muscular dystrophy.

n

WEAKNESS: DIFFERENTIAL DIAGNOSIS

TABLE 5–62

UMN signs Cerebrum, Tumor,

brainstem,

syrinx,

spinal cord multiple sclerosis

(+) Sensory

changes

LMN signs Peripheral nerve Neuropathy

Weakness

UMN signs Anterior horn cell, Amyotrophic lateral

cortical spinal tract sclerosis

(–) Sensory

changes

LMN signs

Anterior horn cell Poliomyelitis

Neuromuscular Myasthenia gravis,

junction Lambert-Eaton syndrome

Pain Polymyositis

Muscle

Painless Myopathy

LMN, lower motor neuron; UMN, upper motor neuronDISORDER

SEPTIC

ENCEPHALOPATHY

CRITICAL ILLNESS

POLYNEUROPATHY

CRITICAL ILLNESS

MYOPATHY

Etiology SIRS SIRS SIRS

Clinical presentation Altered mental status,

an early complication of SIRS

Difficulty weaning from the

ventilator; flaccid limbs

Respiratory weakness;

flaccid limbs

Labs Near normal CSF;

may have normal head

scans, suggesting functional

vs. structural impairment

Normal CK;

muscle biopsy; denerva-

tion atrophy; other labs

altered as with SIRS

Variably elevated; muscle

biopsy; loss of thick

­ filaments; other labs

altered as with SIRS

EDX Motor and sensory NCS:

Axonal loss pattern or

absent

EMG: distal pattern of

decreased recruitment

with or without denerva-

tion and reinnervation,

depending on the time

course

Motor NCS: Low ampli-

tudes throughout

Sensory NCS: Normal

EMG: Small, short,

­ polyphasic MUAPs

with normal or early

­ recruitment; active

­ denervation may be

present

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CRITICAL ILLNESS NEUROMUSCULAR DISEASE (TABLE 5-63)

•

In the ICU patient, various factors including infection, trauma, surgery, chemical exposure, and

­ sepsis can lead to systemic inflammatory response syndrome (SIRS). Single or multiple organ

failure can ensue. Septic encephalopathy and critical illness polyneuropathy (CIP) may result.

Neuromuscular blocking agents and steroids added to this may cause critical illness myopathy

(CIM).

•

SIRS is present if two or more of the following occurs:

– – Body temperature >38°C or <36°C

– – Heart rate >90 bpm

– – Tachypnea:

nn Respiratory rate >20 breaths per minute

nn Hypocapnea PaCO2 <32 torr (<4.3 KPa)

– – White blood cell (WBC) >12,000 or <4,000, or >10% immature neutrophils (bands)

•

CIP is the degradation of neural tissue due to multiple medical complications causing a primar-

ily axonal as well as demyelinating motor and sensory peripheral polyneuropathy.

•

CIM is an acute, generally inflammatory myopathy due to multiple medical complications

causing muscle membrane instability and muscle cell breakdown.

•

Bilateral diffuse weakness predominant in the proximal part of the limbs after improvement

of the acute phase of the critical illness is highly suggestive of critical illness neuromuscular

disease (Table 5–63).

TABLE 5–63 Critical Illness Neuromuscular Disease

CIM, critical illness myopathy; CIP, critical illness polyneuropathy; CK, creatine kinase; CSF, cerebrospinal fluid;

EDX, electrodiagnostic; EMG, ­ electromyography; MUAP, motor unit action potential; NCS, nerve conduction study;

SIRS, ­ systemic inflammatory response syndrome.

Treatment

•

•

Multifactoral and difficult. Treat underlying sepsis and multiorgan failure. Supportive care.

Other disorders of muscle in the differential in the ICU patient: Rhabdomyolysis necrotizing

­ myopathy, disuse myopathy.

•

Other disorders to differentiate from CIP: Spinal cord compression, MND, Guillain–Barre

­ syndrome, myasthenia gravis, myasthenic syndrome.WEAKNESS: DIFFERENTIAL DIAGNOSIS

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CNS DISORDERS

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In CNS disorders, sensory and motor NCSs are normal.

On needle EMG, there is no denervation or reinnervation on the weak limb(s); it reveals normal

spontaneous activity and normal MUAP morphology. On voluntary contraction, however, the

interference pattern is not complete due to decreased activation (i.e., decreased firing fre-

quency); the number of available MUAPs (i.e., recruitment) remains normal.

•

In a segmental spinal cord lesion, muscle innervated below the level of lesion will display the

typical pattern of CNS lesion (i.e., decreased activation). At the segmental level of the lesion,

anterior horn cells may be affected, leading to a neuropathic pattern in muscles innervated by

that segment.

PARANEOPLA