INVITED COMMENTARY

ELECTROPHYSIOLOGICAL CRITERIA OF DEMYELINATION: ITS RELEVANCE TO IMMUNE-MEDIATED NEUROPATHIES

Shin J OH, M.D.

The University of Alabama at Birmingham, The Veterans Affairs Medical Center, Birmingham, Alabama.
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ABSTRACT : *The majority of immune mediated neuropathies are due to segmental demyelination and are treatable with the various immunotherapies. Thus, it is important for clinician to recognize demyelinating neuropathy and confirm it with the electrodiagnostic tests. In this review, we have discussed the electrophysiological hallmarks of demyelination, controversies on conduction block, and electrophysiological characteristics in the various immune mediated neuropathies.*

Key Words: *Guillain-Barre Sydrome, Chronic Inflammatory Demyelinating Polyradiculoneuropathy, Electromyography.*

INTRODUCTION

In demyelinating neuropathy, the diseased process affects the myelin sheath producing primary segmental demyelination and leaving the axon intact (Fig. 1). Segmental demyelination is induced by metabolic damage of Schwann cells or peeling and engulfment by activated inflammatory cells. Myelin is responsible for the conduction of nerve action potentials along the nerve by "saltatory conduction" and, thus, nerve conduction in demyelinating neuropathy is characterized by marked slowing or conduction block (Table 1). Fibrillation or positive sharp waves are either absent or rare in the needle EMG because they are induced predominantly by axonal degeneration. Recovery occurs by the process of remyelination of the shorter intersegments on the demyelinated segment. Once remyelination begins, recovery is usually rapid and complete. The majority of immune mediated neuropathies are due to segmental demyelination and, thus, are treatable with the various immunotherapies. At the University of Alabama at Birmingham (UAB) clinic, we assume that all acquired demyelinating neuropathies are treatable until proven otherwise. Thus, it is important for the clinician to recognize demyelinating neuropathy and confirm it with the electrodiagnostic tests.

Pathologically, segmental demyelination is best seen in the teased nerve fibers (Fig. 2). In the semithin section with osmium tetroxide, demyelinating neuropathy is diagnosed by thinly myelinated fibers representing remyelination and onion-bulb formation, the hallmark of repeated demyelination and remyelination (Figs. 3 and 4). Demyelinated fibers are rarely observed in the semithin section. Scattered epineurial

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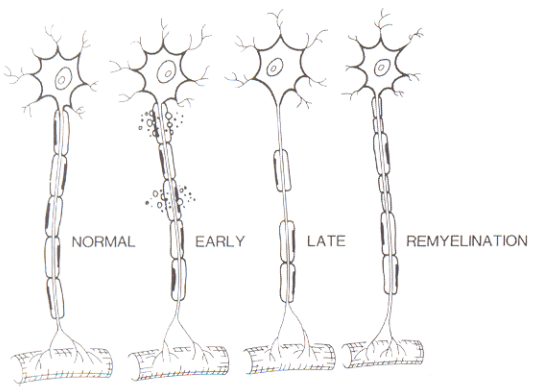

Fig. 1: Mechanism of segmental demyelination and remyelination. Segmental demyelination is induced by metabolic damage of Schwann cells or peeling and engulfment by activated inflammatory cells (early). This process affects the myelin sheath producing primary segmental demyelination and leaving the axon intact (late). Remyelination occurs with myelination over the demyelinated segment.

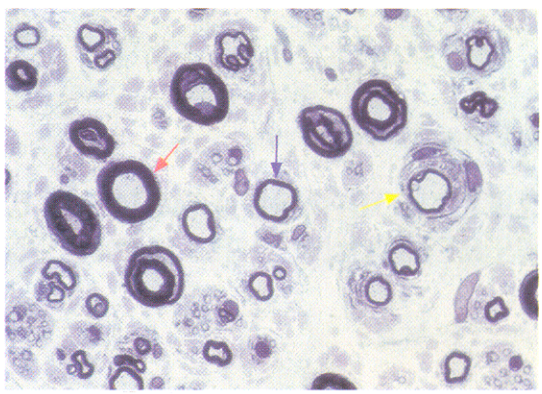

Fig. 3: Thinly myelinated fibers (remyelinated fibers) in CIDP. About 50% of myelinated fibers are remyelinated fibers (blue arrow) characterized by a thin myelin sheath in proportion to axon diameter. Red arrow indicates normal myelinated fibers. Yellow arrow indicates thinly myelinated fibers with two Schwann cell nuclei and tiny onion bulb formation (OBF). Semithin section. Toluidine blue. x 400.

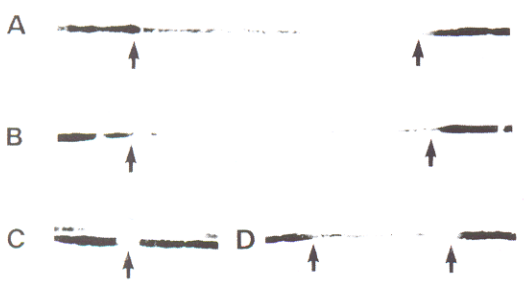

Fig. 2: Teased nerve fibers in the ventral roots of lumbosacral nerves in a case of multifocal motor neuropathy. A and B represent segmental demyelination, and C and D paranodal widening between the arrows (osmium tetroxide, x 100).

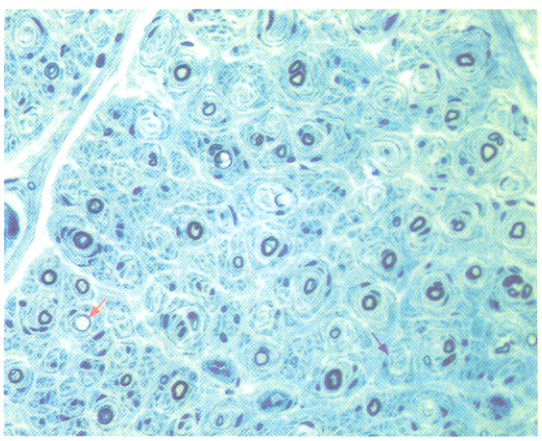

Fig. 4: Onion-bulb-formation (OBF). OBF is recognized by many fine lines around nerve fibers with varying myelin thickness. Red arrow indicates denuded axon (demyelination) and blue arrow, OBF without any recognizable axon. Some fibers with OBF have more than one Schwann cell nucleus. Semithin section. Toluidine blue. x 400.

inflammatory cells are characteristically observed in the inflammatory demyelinating neuropathies such as Guillain-Barre syndrome (GBS) and chronic inflammatory demyelinating polyneuropathy (CIDP) (Fig. 5).

Electrodiagnostic Tests

Segmental demyelination can be recognized now with confidence, by the classical nerve

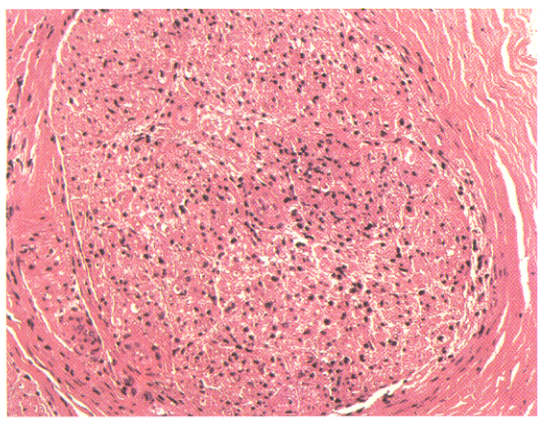

Fig. 5: Epineurial inflammatory cells in CIDP. Many mononuclear inflammatory cells are scattered throughout the endoneurial space. Paraffins section. H & E, x 300.

conduction abnormalities, which are distinctly different from axonal degeneration (Table 2). This has become possible because of the better understanding of the pathophysiology of nerve conduction in demyelinating neuropathies during the past three decades. This is especially true in multifocal motor neuropathy (MMN) or multifocal motor and sensory demyelinating neuropathy (MMSDN) because this is initially recognized by the presence of conduction block.

The hallmarks of nerve conduction abnormalities in segmental demyelination are (1): (1) marked slowing in the nerve conduction velocity (NCV) (2), marked prolongation of terminal, F-wave and H-reflex latencies (3), abnormal temporal dispersion (dispersion phenomenon) and (4) conduction block.

Thus, motor nerve conduction is critical for detection of demyelination in the nerve conduction test because it can detect all of the hallmarks of demyelination in the routine test. On

Table 1: Pathophysiology of Two Types of Peripheral Neuropathy.

Type	Axonal neuropathy	Demyelinating neuropathy
Primary lesion	Axon	Myelin
Pathological process	Axonal degeneration	Demyelination
Pathology by teasing preparation	Myelin ovoids	Segmental demyelination
Regeneration:		
mechanism	Axonal sprouting	Remyelination
speed	Slow	Rapid
Nerve conduction:		
velocity	Mildly slow; above 30 m/sec	Markedly slow; below 30 m/sec
CMAP	Low amplitude	Dispersion; conduction block
Needle EMG:		
Fibrillation & positive sharp wave	(++++)	(-) or (+-)
Fasciculation	Absent	Present in chronic form
Examples:	Arsenic, thallium, gold	Guillain-Barré syndrome
	Alcoholic	CIPD
	Nutritional	Hypertrophic
	Vasculitic	Metachromatic
	Giant axonal	Tomaculous
	Porphyric neuropathy	Leprosy
	Vitamin B12	Diphtheritic
	Diabetic neuropathy	Charcot-Marie-Tooth 1 A
	Uremic neuropathy	

Table 2: Electrophysiological characteristics in axonal degeneration and segmental demyelination.

Characteristic	Axonal Degeneration	Segmental Demyelination
Motor Nerve Conduction (with surfice electrodes)		
Amplitude	↓↓	N or ↓; conduction black
Duration	N	Dispersion phenomenon
Shape	N	N or multiphasic
Terminal latency	N or ↑ (< 150%)	↑↑ (> 150%)[b]
Conduction velocity	N or ↓ (> 60%)	↓↓ (< 80%)
Sensory Nerve Conduction		
With surface electrodes		
Amplitude	↓↓ or often abseant	N, ↓, or absent
Duration	N	↑ or rarely dispersion phenomenon
Shape	N	Rarely multiphasic
Conduction velocity	N or ↓ (> 80%)	↓↓ (< 60%)
With near-nerve needle technique		
Amplitude	↓↓	N or ; conduction black
Duration	N	Prominant dispersion phenomanon
Shape	N	Multiphasic with many components
Conduction velocity	N or ↓ (> 60%)	↓↓ (< 60%)
F-wave	↑ < 150% or absent	↑ > 150% or absent
H-reflex	↑ < 150% or absent	↑ > 150% or absent

↑. Increased ↓. Decreased N. Normal
Percentage of normal means.

the other hand, sensory nerve conduction in the routine test is extremely limited for this purpose because the routine test usually uses the surface recording electrodes; one segment is only tested; and sensory nerve potential is usually absent in the moderate or marked NCV slowing.

In motor conduction, the following findings are typical of segmental demyelination:

1. Markedly slow NCV. Slowing of the NCV depends on the degree of segmental demyelination of large-diameter fibers. NCV slowing by more than 40% below the normal mean (NCV below 60% of normal mean) is indicative of segmental demyelination. Motor NCVs below 35 m/s in the median nerve and below 30 m/s in the peroneal nerve are indicative of segmental demyelination (Fig. 6).

2. Marked prolongation of the terminal latency. The terminal latency is prolonged by more than 50% of the normal mean (terminal latency >150% of normal means). Terminal latencies longer than 6 ms in the median nerve and longer than 10 ms in the peroneal nerve are indicative of segmental demyelination (Fig. 7).

3. Abnormal temporal dispersion or "dispersion phenomenon" (abnormal compound muscle action potential (CMAP) with multiple phases and prolonged duration) (Fig. 8). This is judged in comparison either with the normal CMAP or with the CMAP at the distal site. We prefer the term "dispersion phenomenon" when comparing with the normal CMAP and "abnormal temporal dispersion", when comparing with the CMAP at the distal site. Because abnormal temporal dispersion or dispersion phenomenon represents a

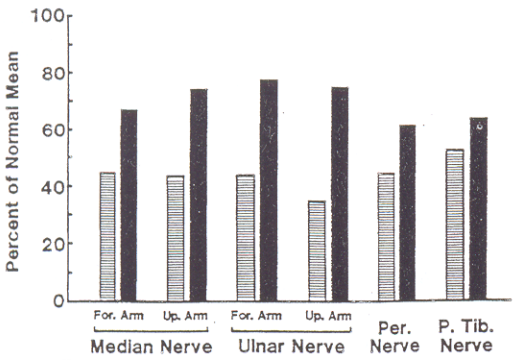

Fig. 6: Comparison of the motor NCVs of axonal degeneration (arsenic neuropathy) and segmental demyelination (CIDP). The hatched bar and solid bar represent segmental demyelination and axonal degeneration, respectively. The normal mean is at 100%, and the lower normal limit at 80% of the mean. With segmental demyelination, the motor NCV's are below 60% of the normal mean, whereas with axonal degeneration they are above 60% of the normal mean.

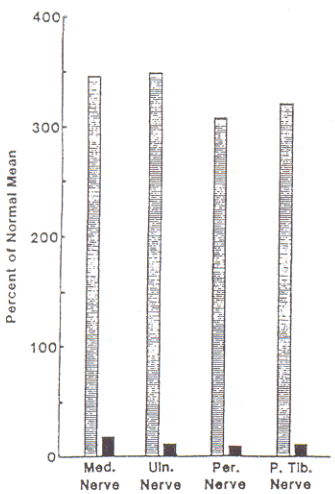

Fig. 7: Comparison of the terminal latency of axonal degeneration (arsenic neuropathy) and segmental demyelination (CIDP). The hatched bar represents mean terminal latencies from 18 cases of CIDP, and the solid bar from eight cases of arsenic neuropathy. The normal mean is 0%. Markedly prolonged terminal latencies are noted in the presence of segmental demyelination, whereas with axonal degeneration terminal latencies are still within normal limits.

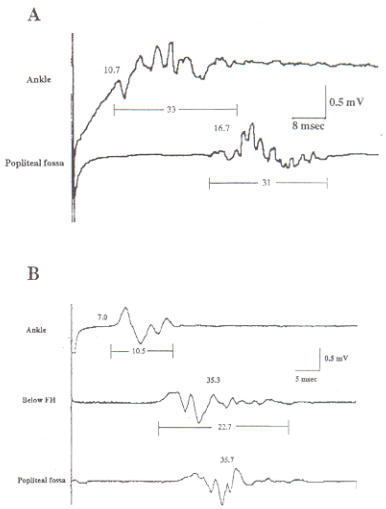

Fig. 8: A. Dispersion phenomenon of the CMAP indicative of demyelination in the posterior tibial motor nerve conduction in a case of chronic sensory demyelinating neuropathy. Dispersion is obvious at ankle as well as at knee. Notice a long duration of CMAP (33 and 31 msec) between two lines. Ankle, Ankle stimulation. Popliteal fossa, popliteal fossa stimulation. Numbers above the CMAP refer to latency (msec) at the ankle and NCV (m/sec) at knee. B. Proximal dispersion phenomenon (temporal dispersion) indicative of focal demyelination between ankle and below-fibular head (FH) in the peroneal motor nerve conduction in a case of chronic inflammatory demyelinating polyneuropathy. Dispersion is obvious at below-fibular head compared with the distal CMAP duration (10.5 msec).

dysynchronization of CMAP due to a wide variation of NCVs of large-diameter fibers, it is typical of segmental demyelination.

4. Conduction block (Fig. 9) . Conduction block is indicative of focal demyelination. The amplitude is somewhat lower with stimulation at more proximal sites in normal subjects. Conduction block is defined as being present when there is a greater than 50-60% reduction of CMAP amplitude and area with normal duration with proximal stimulation as compared with distal stimulation. To document conduction block, stimulation at two sites is needed.

5. Normal or reduced amplitude of the CMAP. Substantial slowing of the NCV in the presence of a normal amplitude is indicative of segmental demyelination. When the amplitude is reduced, this reduction is proportional to the degree of temporal dispersion of the CMAP and

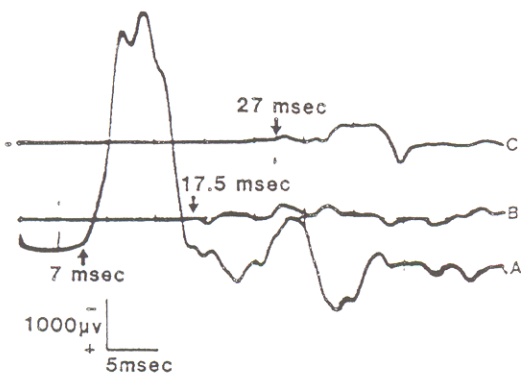

Fig. 9: Conduction block in segmental demyelination. Median motor nerve conduction in a case of CIDP. A, Normal amplitude of the CMAP with wrist stimulation. B, A dramatic reduction in amplitude of the CMAP with elbow stimulation. C, CMAP with axilla stimulation. Conduction block is clearly seen between wrist and elbow stimulation. The dispersion phenomenon is also observed. The motor NCV is 21.9 m/sec over the elbow-wrist segment and 15.8 m/sec over the axilla-elbow segment.

conduction block.

In sensory nerve conduction, the following findings are indicative of segmental demyelination:

1. Marked slowing in NCV. Slowing of the sensory NCV depends on the severity of segmental demyelination. NCV slowing by more than 40% below the normal mean is indicative of segmental demyelination. Sensory NCV below 30 m/s in the median nerve and below 25 m/s in the sural nerve is indicative of segmental demyelination. It is rare to obtain an NCV below 20 m/s with surface electrodes in severe segmental demyelination because the sensory compound nerve action potential (CNAP) is unobtainable due to the smaller amplitude. However, this can be reliably achieved with the near-nerve needle sensory nerve conduction.

2. Dispersion phenomenon or abnormal temporal dispersion. In the sensory nerve conduction, dispersion phenomenon is usually observed, since sensory nerve conduction is tested in one segment in the routine study. On very rare occasions this phenomenon can be documented with the surface electrode, but it is best documented with the near-nerve needle electrode after signal averaging (Fig. 10). When the sensory CNAP is markedly split and lengthened in duration with numerous smaller potential components after the main component, this is termed dispersion phenomenon, which is indicative of segmental demyelination.

3. Conduction block. Since sensory nerve conduction is tested in one segment in the routine nerve conduction study, conduction block cannot be documented by routine sensory nerve conduction. This can only be demonstrated when the sensory fibers are orthodromically stimulated and the recording electrodes are placed in the short segment distal and proximal to the site of block, best seen with the near-nerve needle technique. In practice, because of these technical difficulties, it is almost impossible to demonstrate conduction block in the sensory nerve conduction.

4. Normal or reduced amplitude of the sensory CNAP. Substantial slowing of the NCV in the presence of normal amplitude is indicative of segmental demyelination. When the amplitude

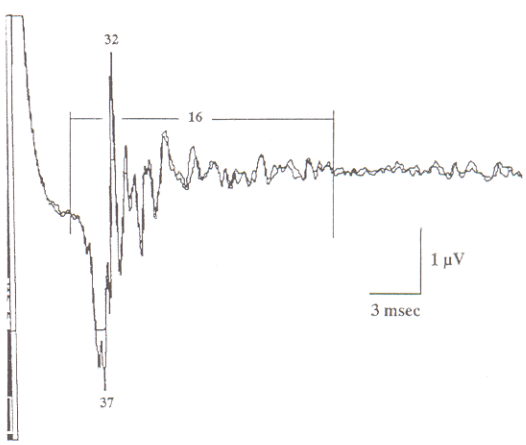

Fig. 10: Dispersion phenomenon in sensory nerve conduction. Dispersion phenomenon of the sensory CNAP indicative of demyelination in the near-nerve needle sensory nerve conduction of medial plantar nerve in a case of Guillain-Barré syndrome. Nerve conduction in the other nerves shows motor neuronopathy without any feature of demyelination. Notice normal maximum and negative peak NCVs (numbers at the positive peak and negative peak) and long duration (16 msec) of the CNAP.

is reduced, it is in proportion to the temporal dispersion and conduction block. With surface electrodes, the sensory CNAP is unobtainable in cases of severe segmental demyelination.

Though there has been uniform agreement on the diagnostic criteria of the NCV and latency for demyelination and temporal dispersion or dispersion phenomenon, there have been controversies about the conduction block. The first controversy is in regard to whether conduction block has to be associated with clinical weakness. Kimura took the view that an electrophysiologic conduction block without clinical weakness does not exist (2). This is easily disproved by our observation that conduction block is present in many chronic sensory demyelinating neuropathy cases and a few sensory GBS patients (3, 4). The second controversy is in regard to what is the best parameter for the detection of conduction block. This study was carried out in the UAB laboratory with the help of Dr. Kuruoğlu, Assistant Editor of this Journal (5). We found that there is no statistically significant difference among the various methods (peak-to-peak amplitude, negative-peak amplitude, total area, and negative area), though the highest detection value was obtained with the total area method. The third controversy is the degree of percentage for the definition of conduction block: 20% to 50%. Our study again showed that 20% is clearly unacceptable because a 41% amplitude reduction was possible in the posterior tibial nerve in normal controls (5). Considering Rhee's conclusion that "area reduction greater than 50% indicates at least some degree of conduction block" on the basis of computer model study (6), we recommended a more than 50% reduction of the CMAP amplitude and area with normal duration as the criterium of conduction block (1). In the recent American Association of Electrodiagnostic Medicine (AAEM) consensus, stricter criteria were used for the peroneal and posterior tibial nerves: 60% reduction of amplitude or area (7). The fourth controversy is the relationship between temporal dispersion (duration) and conduction block. It is well known that temporal dispersion can produce a reduction of amplitude, producing a false conduction block. Our study in normal controls clearly showed that a 30-33 % increase in duration was observed in the negative and total duration respectively in the posterior tibial nerve (5). On the basis of this, the AAEM consensus adopted the duration < 30% increase as the required criterium for definite conduction block (7).

Demyelinating Polyneuropathy

Uniform demyelinating neuropathy is a useful concept in differentiating between hereditary motor sensory neuropathy and acquired chronic demyelinating neuropathy. In uniform demyelinating neuropathy, the NCV is slowed "uniformly" to the same degree over all nerves and nerve segments (Fig. 11) (8, 1). In addition, conduction block or abnormal temporal dispersion is absent because of the uniform demyelination of all fibers. This pattern is classically described as hereditory motor and sensory neuropathy (HMSN) type I Charcot-Marie-Tooth (CMT 1A) and familial neuropathies associated with metachromatic leukodystrophy, globoid leukodystrophy, and Cockayne's syndrome. Exceptions to this rule are hereditary neuropathy to pressure palsy, sex-linked CMT, and Dejerine-Sottas neuropathy (Fig. 12). In acquired demyelinating neuropathies, the non-uniform slowing of NCV over different nerves and nerve segments,

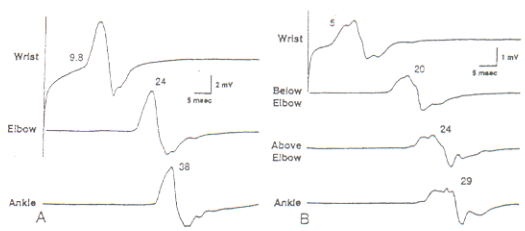

Fig. 11: Uniform demyelination in the CMT 1A. "Uniform slowing" typical of CMT1A. Conduction block and abnormal temporal dispersion are absent. A. Median nerve: NCVs between the segments are "uniformly slow". B. Ulnar nerve: NCVs between the segments are "uniformly slow". Numbers on the left represent the terminal latencies in msec. Numbers on the right represent the NCVs. NCVs over the forearm segment is "uniformly slow" in the median and ulnar nerves.

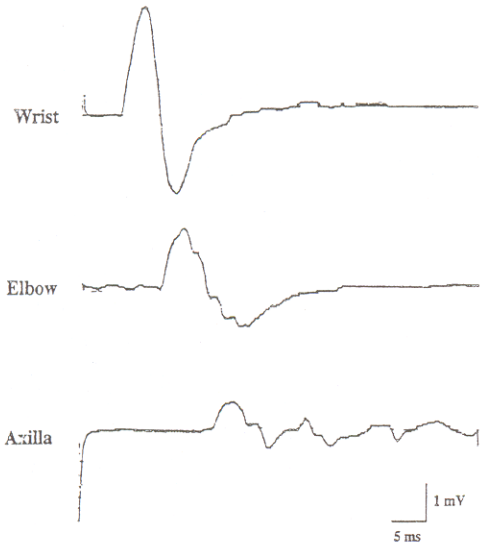

Fig. 12: Non-uniform demyelination in Hereditary neuropathy with liability to pressure palsy. Left median motor nerve conduction. Conduction block is present in the elbow-wrist segment and the axilla-elbow segment. Temporal dispersion is also present in the CMAP with axilla stimulation.

conduction block, and abnormal temporal dispersion are typically observed.

In contrast to acquired axonal neuropathies, most of the acquired demyelinating neuropathies are autoimmune-mediated and treatable. Thus, it is important to recognize these entities. Most of these neuropathies are diagnosed by the clinical constellation and electrophysiological evidences of demyelination. In fact, in some of these entities, the electrophysiological criteria has become an integral diagnostic criterium of demyelination: multifocal motor neuropathy and chronic sensory demyelinating neuropathy. As pure motor demyelinating neuropathies, multifocal motor neuropathy and Guillain-Barré syndrome are listed. Though most CIDP cases are characterized by mixed motor-sensory neuropathy, there are subgroups of patients who have pure motor demyelinating neuropathy. As pure sensory demyelinating neuropathies, acute demyelinating neuropathy exists as a sensory variant of GBS (4) and chronic sensory demyelinating neuropathy (CSDN) as a sensory variant of CIDP (3). Multifocal motor sensory demyelinating neuropathy (MMSDN) also exists as an intermediate link between CIDP and MMN (9, 10). We have observed one case of multifocal sensory demyelinating neuropathy (MSDN): pure sensory neuropathy involving the left upper extremity.

GBS, acute inflammatory demyelinating neuropathy, is characterized clinically by ascending areflexic and predominantly motor weakness with peak deficit within 4 weeks of onset, by a high CSF protein in almost all cases, antecedent infection in 75% of cases, common facial nerve involvement and respiratory failure in 20% of cases. In GBS, nerve conduction abnormalities are present in almost all cases and there is evidence of demyelination in 80% of cases (1). Demyelination in GBS can be either expressed as marked NCV slowing, temporal dispersion, or conduction block. It is important to remember that in 20% of cases, NCV is minimally slow, in the range of "axonal neuropathy" due to mild demyelination

Sensory GBS, a sensory variant of GBS, which we have recently reported, is identical with the classical GBS except for pure sensory symptoms (4). In this again, the motor nerve conduction was critical in recognizing demyelination and the most common abnormality was the prolonged terminal latency to the demyelinating range. In one case, the near-nerve

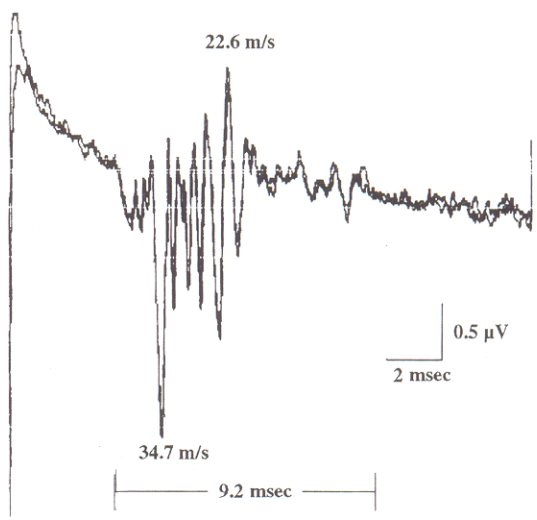

Fig. 13: Dispersion phenomenon in Sensory GBS. Dispersion phenomenon indicative of demyelinating neuropathy in the near-nerve needle sensory nerve conduction of the I-II interdigital nerve in a case of sensory GBS with normal routine nerve conduction. Markedly slow (22.6 m/s) negative-peak NCV and long duration (9.2 msec) of CNAP are indicative of demyelination.

sensory nerve conduction was used to document demyelination (Fig. 13).

CIDP, chronic form of GBS, is characterized by subacute or chronic polyneuropathy with progression over a more than 4 weeks period and high CSF protein. Other clinical differences are listed in Table 3. In CIDP, the most helpful objective diagnostic finding is the typical pattern of nonuniform demyelinating neuropathy (1). Without this pattern, it is impossible to make the diagnosis of CIDP. The nerve conduction test in this disorder shows findings typical of extensive segmental demyelination: marked slowing in NCV, conduction block, and dispersion phenomenon. The nerve conduction abnormalities are diffuse, involving the distal and proximal segments of the nerve. All nerves are affected in this disorder, although the degree of abnormality varies from nerve to nerve. Motor nerve conduction is universally abnormal in CIDP, showing evidence of demyelination. Sensory CNAPs are either absent or low in amplitude, and the F-wave and H-reflexes are either absent or markedly prolonged in latencies. Compared with GBS, CIDP is characterized by more typical and severe evidence of demyelination (Fig. 14).

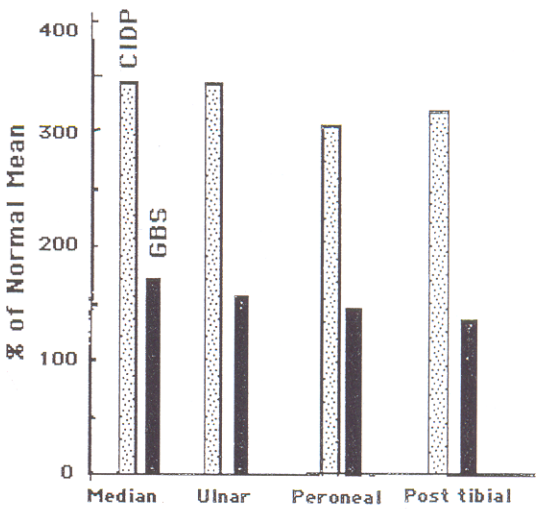

Fig. 14: Comparison of the terminal latency in GBS and CIDP. The dotted bar represents mean terminal latencies from 18 cases of CIDP, and the solid bar from 28 cases of GBS. The normal mean is 0%. Markedly prolonged terminal latencies are noted in CIDP, with moderately prolonged terminal latencies in GBS.

Chronic sensory demyelinating neuropathy (CSDN) is an entity recently described by us with Dr. Kuruoğlu as a coauthor and is characterized by subacute or chronic progression over months or years, pure sensory neuropathy (no motor weakness), high spinal fluid protein in most cases, electrophysiological evidence of demyelination affecting motor and sensory nerve fibers, demyelination on sural nerve biopsy (Fig. 15), and good response to immunotherapy in the progressive phase of disease (3). In our experience, CSDN is the most common treatable form of neuropathy among the sensory neuropathies. We believe that this entity represents a CIDP presenting as pure sensory neuropathy. The most important diagnostic test in CSDN is the motor nerve conduction study, which is usually the first objective clue

Table 3: Differences between the Guillain-Barre syndrome and chronic inflammatory demyelinating polyneuropathy.

Features	Guillain-Barré Syndrome	CIDP
Onset	Acute; maximum neurological Deficit in 2-3 weeks	Subacute; maximum neurological deficit in 1-12 months
Antecedent infection	Present in 70%	Absent
Cranial nerve deficit	Common	Rare
Respiratory muscle	Common	Rare
NCV	Normal or slightly slow in 20%; markedly slow in 80%	Markedly slow
Response to steroids	Not proved	Positive
Relapse	Rare	Common

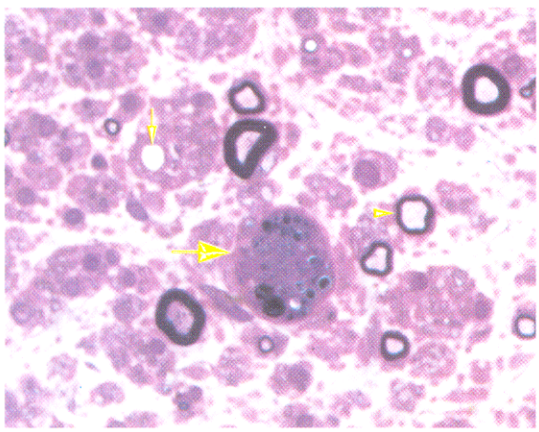

Fig. 15: Sural nerve biopsy in CSDN. Moderate loss of myelinated fibers. Arrow indicates one denuded axon. Arrowhead indicates one of many thinly myelinated fibers here. Larger arrow indicates a lipid-laden macrophage. Semithin section. Toluidine blue & basic fuchsin. x 1000.

suggestive of demyelinating neuropathy. In some patients, the near-nerve needle sensory nerve conduction study is needed to make a definite diagnosis.

MMN has emerged as a distinct entity in recent years and can be confused with amyotrophic lateral sclerosis (ALS) because of pure motor involvement, frequent fasciculation, preserved or brisk reflexes, and frequent onset of weakness in the arms (11-13). This neuropathy is characterized clinically by pure motor mononeuropathy multiplex, immunologically by a strong association with the anti monosialo ganglioside (GM1) antibody, and therapeutically by a lack of steroid response but a fair response to cyclophosphamide and IVIg treatment. Our studies with the help of Dr. Odabaşı in Turkey, have documented pathologically that MMN is inflammatory demyelinating polyradiculoneuropathy, not much different from those in CIDP (14). The electrophysiological hallmark of this disorder is multifocal motor conduction block with sparing of sensory nerve conduction (15, 9). Many claimed that without conduction block one can not make the diagnosis of this disorder. According to our experience, one can make the diagnosis of MMN without motor conduction block, and minor sensory nerve conduction abnormalities are not uncommon.

MMSDN resembles MMN because it is a multifocal neuropathy, involves the arms predominantly , and shows demyelination in the NCS (16, 9,10, 17). However, it is different from MMN because of the presence of sensory deficits, sensory nerve conduction abnormalities, good response to immunotherapies, and absence of GM1 antibody. This entity is characterized by subacute or chronic motor-sensory mononeuropathy multiplex, high CSF protein in 60% of patients, demyelination in the nerve biopsy, and good response to the immunotherapies. One case we investigated with the help of Dr. Demirci in Turkey showed that MMSDN is an inflammatory demyelinating polyradiculoneuropathy, identical with those in CIDP. Electrophysiological characteristics of this disorder are evidence of demyelination in multifocal nerves and frequent sensory nerve conduction abnormalities.

In one case of multifocal sensory demyelinating neuropathy (MSDN), the routine NCS showed essentially normal findings except for a low sensory CNAP amplitude in the left median and ulnar nerves (18). Near-nerve needle study in the ulnar nerve, which was performed by Dr. Demirci in Turkey, showed "conduction block" and temporal dispersion in the above-elbow and axilla segment, confirming unequivocally demyelinating neuropathy. This patient had a two-year-history of proprioception loss in the left hand.

Myelin-associated glycoprotein positive (MAG) neuropathy has distinct features: predominantly sensory neuropathy, IgM paraprotein, high CSF protein, and unsatisfactory response to immunotherapies (19, 20). Nerve conduction studies show findings typical of demyelinating neuropathy. The most conspicuous finding in MAG-positive neuropathy is a disproportionate distal slowing of motor conduction expressed by markedly prolonged terminal latency in comparison to the degree of slow NCV in the proximal segment of nerves.

Correspondence to: Shin J Oh, M.D.
The University of Alabama at Birmingham, The Veterans Affairs Medical Center.
BIRMINGHAM - ALABAMA

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