What Are the Electrodiagnostic Criteria for CIDP?

Ken Gorson, MD

Case Presentation

- 37-year-old woman with “trouble walking”
- Slowly progressive over 4 months
- Unsteady gait–poor balance, tripping
- Painless symmetrical leg weakness
- Arms and hands unaffected
- Numb toes and feet

Case Presentation

- History of schizoaffective disease
  - Repeated prior psychiatry hospitalizations for “disordered thinking”
  - Stable with Tegretol, monthly ECT
  - No recent psychotic episodes
- ESRD for 3 years
  - Toxic nephropathy due to chronic lithium treatment
  - Hemodialysis for 6 months

Examination

- Cranial nerves normal
- Grip reduced to 22 kg
- Hand intrinsics 4+/5
- TA: 4/5 bilaterally
- Proximal strength normal
- +2 DTR in arms; areflexic in legs
- Vibration, touch impaired to mid-leg
- Romberg sign

Electrodiagnostic Evaluation

MOTOR NERVE CONDUCTION

Nerve and Stimulation Site	Dist mm	Lat ms	Amp mV	Dur ms	Area mVms	Segment	Lat Diff ms	CV m/s
Peroneal.R to Extensor digitorum brevis.R								
Ankle	80	5.5	**1.2**	7.4	12.9	Extensor digitorum brevis-Ankle	5.5	
Fibula (head)	280	12.6	1.1	8.7	12.5	Ankle-Fibula (head)	7.1	39
Popliteal fossa	100	15.2	0.9	7.8	11.7	Fibula (head)-Popliteal fossa	2.6	**38**
Tibial.R to Abductor hallucis.R								
Ankle	80	6.2	**2.5**	5.3	11.8	Abductor hallucis-Ankle	6.2	
Popliteal fossa	370	16.4	2.1	8.1	2.8	Ankle-Popliteal fossa	10.2	**36**
Median.R to Abductor pollicis brevis.R								
Wrist	70	4.3	**3.7**	5.3	13.3	Abductor pollicis brevis-Wrist	4.3	
Elbow	210	8.9	3.3	5.5	13.1	Wrist-Elbow	4.6	**46**
Ulnar.R to Abductor digiti minimi (manus).R								
Wrist	70	3.0	**5.4**	6.6	24.1	Abductor digiti minimi (manus)-Wrist	3.0	
Below elbow	200	7.3	5.1	6.8	23.3	Wrist-Below elbow	4.3	**47**
Above elbow	100	9.4	4.8	6.9	23.6	Below elbow-Above elbow	2.1	48

Personal case of Jon Katz, MD

Electrodiagnostic Evaluation (cont'd)

F-WAVE

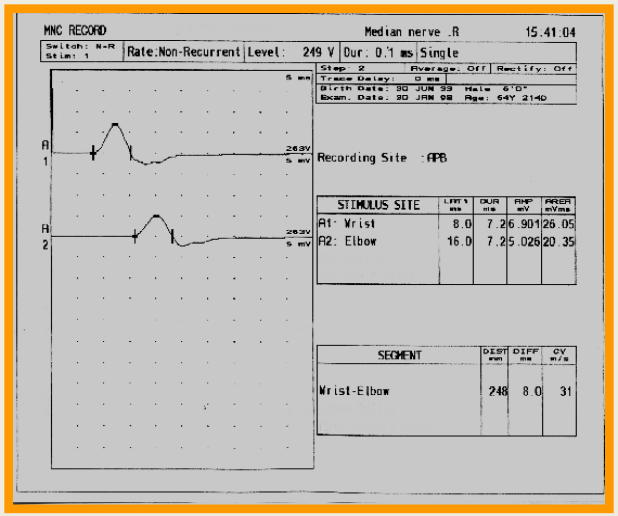
Nerve	M-Lat ms	F-Lat ms
Peroneal.R	15.2	**59**
Tibial.R	16.4	**61**
Median.R	8.9	**32**
Ulnar.R	9.4	31

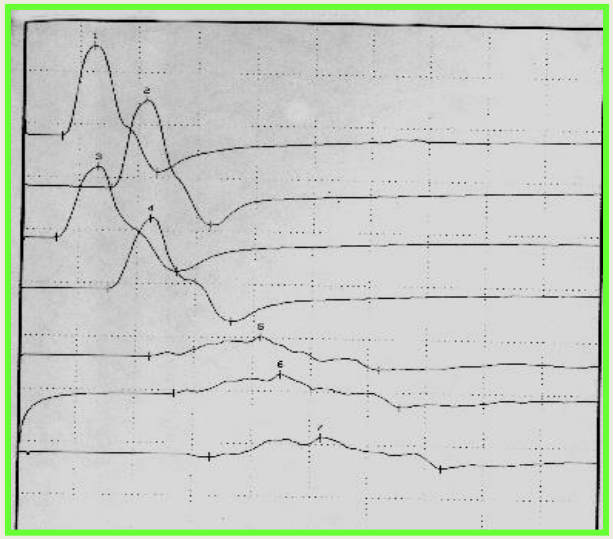
SENSORY NERVE CONDUCTION

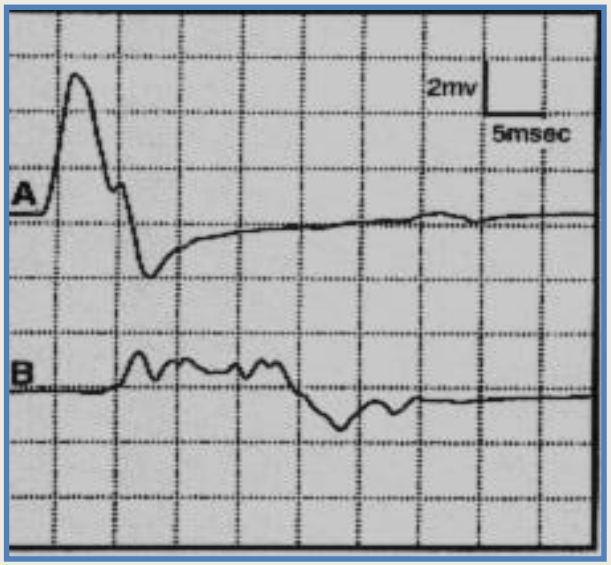
Nerve and Stimulation Site	Dist mm	Onset Lat ms	Peak Lat ms	*Amp* mV	Segment	Lat Diff ms	CV m/s
Superficial peroneal.R to Ankle.R							
Lower leg		NR	NR	**NR**	Ankle-Lower leg		
Sural.R to Ankle.R							
Lower leg		NR	NR	**NR**	Ankle-Lower leg		
Median.R to Digit II (index finger).R							
Wrist	130	2.5	3.2	**8.3**	Digit II (index finger)-Wrist	2.5	52
Ulnar.R to Digit V (little finger).R(manus).R							
Wrist	110	2.3	3.1	**9.2**	Digit V (little finger)-Wrist	2.3	47

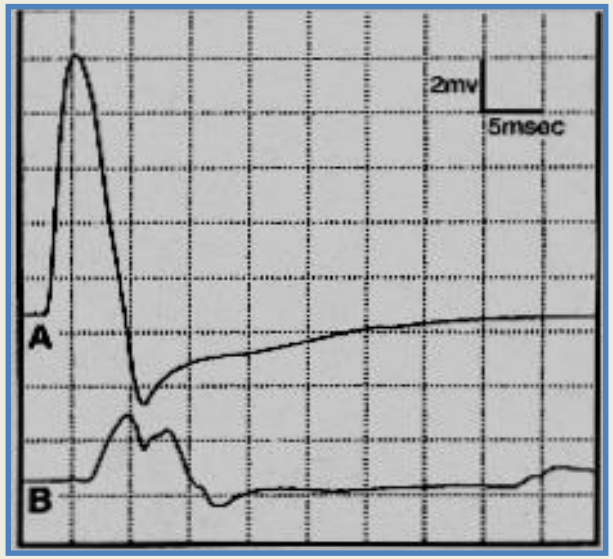
Personal case of Jon Katz, MD

Types of Acquired Demyelination









Personal case of Jon Katz, MD

Gorson's Pearls: Diagnostic Criteria for CIDP

- They exist
- They are underutilized
- They are important to use to guide therapy and prognosis
- Consist of:
  - Clinical criteria—history and examination by qualified neurologist
  - EMG criteria—by qualified electromyographer
- When criteria are not used, there are diagnostic and treatment misadventures

Ken Gorson, Personal Experience

CIDP: The Early Electrodiagnostic Criteria

- Albers and Kelly, 1989

  <u>At least 3 of the following:</u>

  CV < 75% the lower limit of normal (≥ 2 nerves)

  DL > 125% the ULN (≥ 2 nerves)

  F response > 125% ULN (≥ 1 nerve)

  Abnormal Temporal Dispersion/Conduction Block >30%

Albers JW, Kelly JJ Jr. *Muscle Nerve*. 1989; 12(6):435-51.

CIDP: AAN Electrodiagnostic Criteria (1991)

- **Prolonged or absent F-responses**
  - > 120% upper limit of normal with normal distal amplitude
  - 2 or more nerves
- **Prolonged distal motor latencies**
  - > 120% upper limit of normal with normal distal amplitude
  - 2 or more nerves
- **Slowing of conduction velocities**
  - < 80% of lower limit of normal with normal distal amplitude
  - 2 or more nerves
- **Conduction block**
  - > 20% amplitude drop between proximal and distal sites
  - 1 or more nerves

These are strict research criteria (high specificity, low sensitivity)

Research criteria for diagnosis of chronic inflammatory demyelinating polyneuropathy (CIDP). *Neurology*. 1991;41(5):617-618.

Koski Criteria

Patients with a chronic polyneuropathy, progressive for at least 8 weeks, would be classified as having CIDP if:

- No serum paraprotein and
- No documented genetic abnormality

AND EITHER

a) Symmetric onset or symmetric exam, and

ii. Weakness in all 4 limbs, and

iii. At least one limb with proximal weakness

b) At least 75% of motor nerves tested had recorded response AND one of the following conditions is satisfied:

i. More than 50% of the motor nerves tested had abnormal DL or

ii. More than 50% of the motor nerves tested had abnormal CV or

iii. More than 50% of the motor nerves tested had F-latency or

Koski CL, et al. *J Neur Sci*. 2009;277:1-8.

Sensitivity and Specificity of Koski Criteria

Sensitivity and specificity of the proposed rule in the derivation and validation samples

Sample	Sensitivity[a]	Specificity[b]
Derivation sample (n=150)	57/58 (98%)	87/92 (95%)
Validation sample (n=117)	40/48 (83%)	67/69 (97%)

Sensitivity and specificity of previously published diagnostic criteria, as applied to the current study sample (n=267)

Diagnostic criteria	Sensitivity[a] (based on 106 consensus CIDP cases)	Specificity[b] (based on 161 consensus non-CIDP cases)
Barohn et al. 1988[c]	27%	94%
AAA Ad Hoc Subcommittee, 1991[d]	11%	98%
INCAT, 2001[e]	64%	75%
EFNS, 2005[9][f]	34%	99%

Accepted EFNS/PNS Electrodiagnostic Criteria for CIDP[1]

≥ 1 of the following demyelinating parameters are necessary:

- ≥50% prolongation of motor distal latency above ULN in 2 nerves
- ≥30% reduction of motor conduction velocity below LLN in 2 nerves
- ≥20% prolongation of F-wave latency above ULN in 2 nerves, or >50% if amplitude of distal negative peak CMAP is <80% of LLN
- Absence of F-waves in 2 nerves, if nerves have amplitudes of distal negative peak CMAPs ≥20% of LLN, plus ≥1 other demyelinating parameter in ≥1 other nerve
- Partial motor conduction block: ≥50% amplitude ↓of proximal negative peak CMAP relative to distal, if distal negative peak CMAP ≥20% of LLN, in 2 nerves, or in 1 nerve plus ≥1 other demyelinating parameter in ≥1 other nerve
- Abnormal temporal dispersion: >30% duration ↑ between proximal and distal negative peak CMAP in ≥2 nerves
- Distal CMAP duration (interval between onset of the first negative peak and return to baseline of the last negative peak) ↑ in ≥1 nerve (median ≥6.6 ms, ulnar ≥6.7 ms, peroneal ≥7.6 ms, tibial ≥8.8 ms) plus ≥1 other demyelinating parameter in ≥1 other nerve

1. Joint Task Force for EFNS and PNS. *J Peripher Nerv Syst*. 2010;15:1-9.

Shortcomings of Electrodiagnostic Studies in CIDP

Patients may accrue substantial axonal loss with absent or greatly reduced motor potentials; nerve conduction abnormalities may not fulfill criteria for demyelination

Patients with pure sensory variant may have absent sensory nerve activation potential (SNAPs) and relatively normal motor potentials

A few patients have electrodiagnostic abnormalities limited to proximal motor nerve segments that are detected only with motor root or Erb's point stimulation

In clinical practice, a minority of patients do not fulfill accepted EMG criteria, but still may benefit from treatment; CSF analysis and nerve biopsy may be helpful in selected cases to confirm diagnosis

Ken Gorson, personal experience

REVIEW OF THE EVOLUTION OF ELECTRODIAGNOSTIC CRITERIA FOR CHRONIC INFLAMMATORY DEMYELINATING POLYRADICOLONEUROPATHY

MARK B. BROMBERG, MD, PhD

ABSTRACT: Chronic inflammatory demyelinating polyradicoloneuropathy (CIDP) is a treatable form of neuropathy. Efforts to devise sets of electrodiagnostic (nerve conduction) criteria to distinguish primary demyelination from primary axonal neuropathies have been elusive, and at least 16 criteria have been proposed. Modifications to criteria frequently represent minor changes based on applying a set to a small number of patients with the clinical diagnosis of CIDP, whereas others are based on physiological changes related to demyelination and other pathophysiological features. The various modifications continue to result in limited sensitivity, likely related to the wide range of nerve conduction abnormalities among CIDP patients. Although some sets are appropriate for formal clinical drug trials, their complexity makes them difficult to apply in the clinic or electromyography laboratory. This study considers the evolution of the criteria, discusses their limitations, and ends with a simplified set of guidelines that can be applied in the clinic or laboratory.

Bromberg M. *Muscle Nerve*. 2001;43:780-794.

Conclusions

- There are too many criteria
  - Many designed for research not clinics
- Often not applied rigorously or consistently
- Our practice favors EFNS or Koski, et al with similar sensitivity (80%) and specificity (96%)
- Need to educate our colleagues