

# What Are the Electrodiagnostic Criteria for CIDP?

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# 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 intrinsic 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    |
|--|---------|--------|------------|--------|-----------|--------------------------------------|-------------|-----------|
| <b>Peroneal.R to Extensor digitorum brevis.R</b>   |         |        |            |        |           |                                      |             |           |
| Ankle  | 80      | 5.5    | <b>1.2</b> | 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         | <b>38</b> |
| <b>Tibial.R to Abductor hallucis.R</b>             |         |        |            |        |           |                                      |             |           |
| Ankle  | 80      | 6.2    | <b>2.5</b> | 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        | <b>36</b> |
| <b>Median.R to Abductor pollicis brevis.R</b>      |         |        |            |        |           |                                      |             |           |
| Wrist  | 70      | 4.3    | <b>3.7</b> | 5.3    | 13.3      | Abductor pollicis brevis-Wrist       | 4.3         |           |
| Elbow  | 210     | 8.9    | 3.3        | 5.5    | 13.1      | Wrist-Elbow                          | 4.6         | <b>46</b> |
| <b>Ulnar.R to Abductor digiti minimi (manus).R</b> |         |        |            |        |           |                                      |             |           |
| Wrist  | 70      | 3.0    | <b>5.4</b> | 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         | <b>47</b> |
| Above elbow  | 100     | 9.4    | 4.8        | 6.9    | 23.6      | Below elbow-Above elbow              | 2.1         | 48        |

# Electrodiagnostic Evaluation (cont'd)

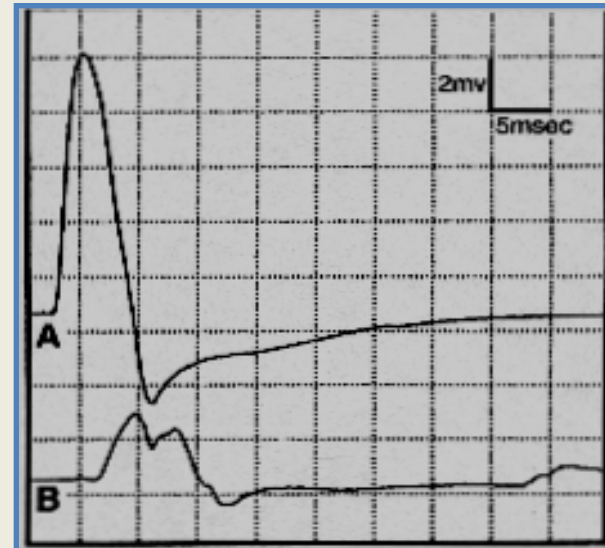
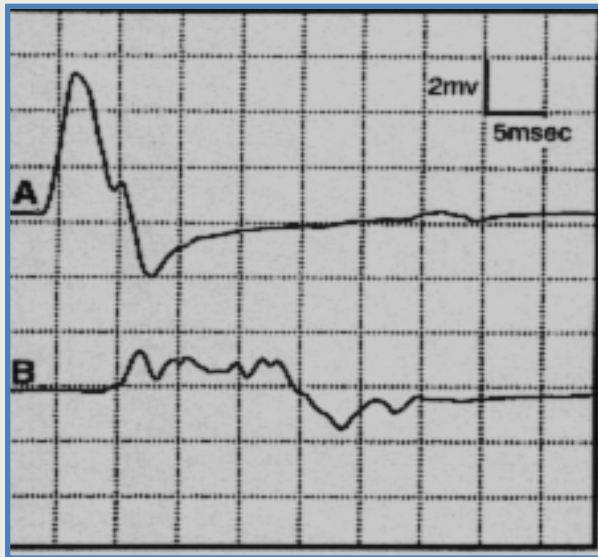
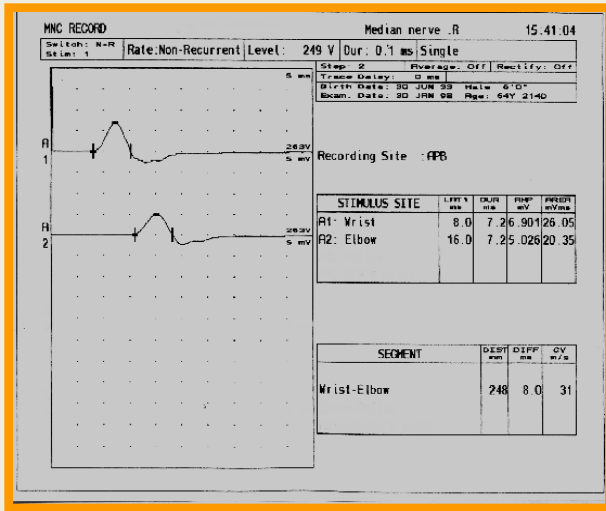
## F-WAVE

| Nerve      | M-Lat<br>ms | F-Lat<br>ms |
|------------|-------------|-------------|
| Peroneal.R | 15.2        | <b>59</b>   |
| Tibial.R   | 16.4        | <b>61</b>   |
| Median.R   | 8.9         | <b>32</b>   |
| 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 |
|--|---------|--------------|-------------|------------|-------------------------------|-------------|--------|
| <b>Superficial peroneal.R to Ankle.R</b>             |         |              |             |            |                               |             |        |
| Lower leg  |         | NR           | NR          | <b>NR</b>  | Ankle-Lower leg               |             |        |
| <b>Sural.R to Ankle.R</b>                            |         |              |             |            |                               |             |        |
| Lower leg  |         | NR           | NR          | <b>NR</b>  | Ankle-Lower leg               |             |        |
| <b>Median.R to Digit II (index finger).R</b>         |         |              |             |            |                               |             |        |
| Wrist  | 130     | 2.5          | 3.2         | <b>8.3</b> | Digit II (index finger)-Wrist | 2.5         | 52     |
| <b>Ulnar.R to Digit V (little finger).R(manus).R</b> |         |              |             |            |                               |             |        |
| Wrist  | 110     | 2.3          | 3.1         | <b>9.2</b> | Digit V (little finger)-Wrist | 2.3         | 47     |

# Types of Acquired Demyelination



# 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



# CIDP: The Early Electrodiagnostic Criteria

- Albers and Kelly, 1989

At least 3 of the following:

CV < 75% the lower limit of normal (  $\geq 2$  nerves)

DL > 125% the ULN (  $\geq 2$  nerves)

F response > 125% ULN (  $\geq 1$  nerve)

Abnormal Temporal Dispersion/Conduction Block  
>30%

# 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)

# 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

# Sensitivity and Specificity of Koski Criteria

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

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

## **≥ 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

# Shortcomings of Electrodiagnostic Studies in CIDP

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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

# REVIEW OF THE EVOLUTION OF ELECTRODIAGNOSTIC CRITERIA FOR CHRONIC INFLAMMATORY DEMYELINATING POLYRADICULONEUROPATHY

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**ABSTRACT:** Chronic inflammatory demyelinating polyradiculoneuropathy (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.

# 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