Electrophysiologic Findings in Multifocal Motor Neuropathy
Multifocal Motor Neuropathy

- Multifocal motor neuropathy (MMN) is a rare asymmetric motor neuropathy without major sensory loss that affects mainly distal extremities\(^1\)

- Diagnosis of MMN is based on clinical and electrodiagnostic criteria\(^2\)

- Here we explore electrophysiologic findings of MMN in addition to the classical electrophysiological conduction block (CB), considered the defining hallmark of MMN\(^1,2\)

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Electrophysiologic Analysis in MMN Patients (I)\(^1\)

- Electrophysiologic findings in MMN were assessed in a retrospective analysis of patients who met a clinically-based diagnosis of MMN
  - Inclusion criteria, designed to find patients with a pure motor involvement in multiple nerves:
    - Age \(\geq 20\) years at onset
    - Weakness in the distribution of \(\geq 2\) peripheral individual motor nerves
  - Exclusion criteria, designed to ensure that patients with ALS and MADSAM were not included:
    - Facial weakness, bulbar weakness, respiratory difficulty, upper motor neuron signs, weakness not localized to individual peripheral nerves, no longer met the study criteria after a follow-up period of at least one year
    - Only minimal sensory involvement was permitted

ALS, amyotrophic lateral sclerosis; MADSAM, multifocal acquired demyelinating sensory and motor neuropathy

Electrophysiologic Analysis in MMN Patients (II)¹

- A total of 16 patients met the clinical criteria for inclusion

<table>
<thead>
<tr>
<th>Clinical characteristic</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex, N</td>
<td>16</td>
</tr>
<tr>
<td>Male</td>
<td>12</td>
</tr>
<tr>
<td>Female</td>
<td>4</td>
</tr>
<tr>
<td>Average age, years</td>
<td>48.1</td>
</tr>
<tr>
<td>Average duration of symptoms, years</td>
<td>7.6</td>
</tr>
</tbody>
</table>

Electrophysiologic Findings in MMN

- Of the 16 MMN patients defined clinically:
  - 31% had evidence of CB in at least one nerve
  - 44% had evidence of temporal dispersion in at least one nerve
  - 94% had other demyelinating features with superimposed axonal degeneration

<table>
<thead>
<tr>
<th>Total patients, N</th>
<th>CB</th>
<th>Possible CB</th>
<th>Temporal dispersion</th>
<th>Slowing conduction velocity</th>
<th>Prolonged distal latency</th>
<th>Prolonged F wave</th>
<th>Pure axonal involvement</th>
</tr>
</thead>
<tbody>
<tr>
<td>16</td>
<td>5</td>
<td>15</td>
<td>15</td>
<td>5</td>
<td>9</td>
<td>8</td>
<td>13</td>
</tr>
</tbody>
</table>

- The ulnar nerve was affected most often in nerve conduction studies
  - Segmental demyelination occurred in 7 proximal, 2 intermediate and 8 distal ulna sites

Conduction Block

- CB is defined as a reduction of compound muscle action potential amplitude on proximal compared with distal nerve stimulation\(^1\)

- In MMN, CB does not affect sensory fibers and can happen at non-compressible sites along the nerve axons\(^2\)

CB on a motor nerve conduction study stimulated at
(A) wrist
(B) elbow
showing an amplitude reduction of 75% and area reduction of 69%. Duration of the negative peak increased by 23%\(^3\)

Conduction Block in MMN

- CB was reported in 5/16 patients
- Other abnormalities were also found in these patients, as follows:

<table>
<thead>
<tr>
<th>Possible CB</th>
<th>Temporal dispersion</th>
<th>Slowing conduction velocity</th>
<th>Prolonged distal latency</th>
<th>Prolonged F wave</th>
</tr>
</thead>
<tbody>
<tr>
<td>3</td>
<td>3</td>
<td>3</td>
<td>2</td>
<td>1</td>
</tr>
</tbody>
</table>

- All patients with CB also had low distal motor amplitudes
- Overall CB was less common than other features

MMN Without Conduction Block

• Patients without CB presented the following features:

<table>
<thead>
<tr>
<th>Possible CB</th>
<th>Temporal dispersion</th>
<th>Slowing conduction velocity</th>
<th>Prolonged distal latency</th>
<th>Prolonged F wave</th>
</tr>
</thead>
<tbody>
<tr>
<td>6</td>
<td>4</td>
<td>10</td>
<td>4</td>
<td>7</td>
</tr>
</tbody>
</table>

• MMN without CB can be due to:
  – Difficulty in confirming CB due to the proximal location of the CB
  – Activity-dependence of these blocks

• CB is not specific for MMN as it can be seen with other types of inflammatory neuropathies, such as CIDP and MADSAM

CIDP, chronic inflammatory demyelinating polyneuropathy; MADSAM, multifocal acquired demyelinating sensory and motor neuropathy

Other Electrophysiologic Features of MMN

- Other electrophysiologic features of MMN can include:¹

- Pure axonal features have also been reported in MMN²,³
  - Clinically defined MMN with no demyelinating features has been referred to as multifocal acquired motor axonopathy, or MAMA

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Axonal Features in MMN

- Motor amplitudes were reduced on distal stimulation in at least one nerve in 14 patients
  - Of these, 8 patients had at least one nerve with pure axonal features

- Lower-extremity weakness was more commonly associated with axonal features in the study of 16 MMN patients, and was seen in:

  - All nerves with CB
  - All nerves with prolonged F waves
  - 9/11 nerves with possible CB
  - 5/10 nerves with temporal dispersion

Response was classed as improvement of at least one Medical Research Council score grade in any muscle group or 30% improvement in handgrip dynamometry.

Response to immunotherapy was variable in patients with distal lower motor neuron weakness without CB.
IVIG Efficacy for MMN With or Without CB

- IVIG is a standard treatment for MMN\(^1\)
- IVIG has been shown to be effective in MMN with and without CB\(^2\)

![IVIG response graph]

- Patients, %
  - Katz et al\(^2\): 2/3
  - Delmont et al\(^3\): 14/20

Although considered an electrophysiological hallmark of MMN, some MMN patients have been reported to have a typical clinical presentation and good response to IVIG even when CB is not found.

Diagnostic criteria for MMN requiring CB may therefore result in underdiagnoses.

Other electrophysiologic features that appear associated with MMN include:

- Prolonged distal latencies
- Temporal dispersion
- Slow conduction velocity
- Delayed or absent F-waves

Awareness of this spectrum of features in MMN is important to increase recognition of this neuropathy.